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ABSTRACT

Intended for physical educators, this manual provides guidelines for providing safe and effective physical activity programs for children with Marfan syndrome, a congenital condition involving the connective tissues and the probable cause of sudden death by heart failure of some young competitive athletes in recent cases. The manual includes information regarding: Public Law 94-142, the Education for All Handicapped Children Act; physical characteristics associated with the disorder and resultant physical needs; potential psychosocial implications; the establishment of educational goals to address these implications; and competencies and/or sensitivities needed by involved professionals. Suggested guidelines for physical activity, instructional methods and strategies, and approved curricular activities are included. In addition, contraindicated activities are identified as well as implications for safety and effectiveness in physical activity programs. The importance of applying these guidelines only under the supervision of the treating physician is stressed. Additional features of the manual include a list of Advisory Panel members of the National Marfan Foundation, a list of organizational resources, and a glossary. Contains 78 references. (DB)

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THE MARFAN SYNDROME

Physical Activity Guidelines For Physical Educators, Coaches And Physicians

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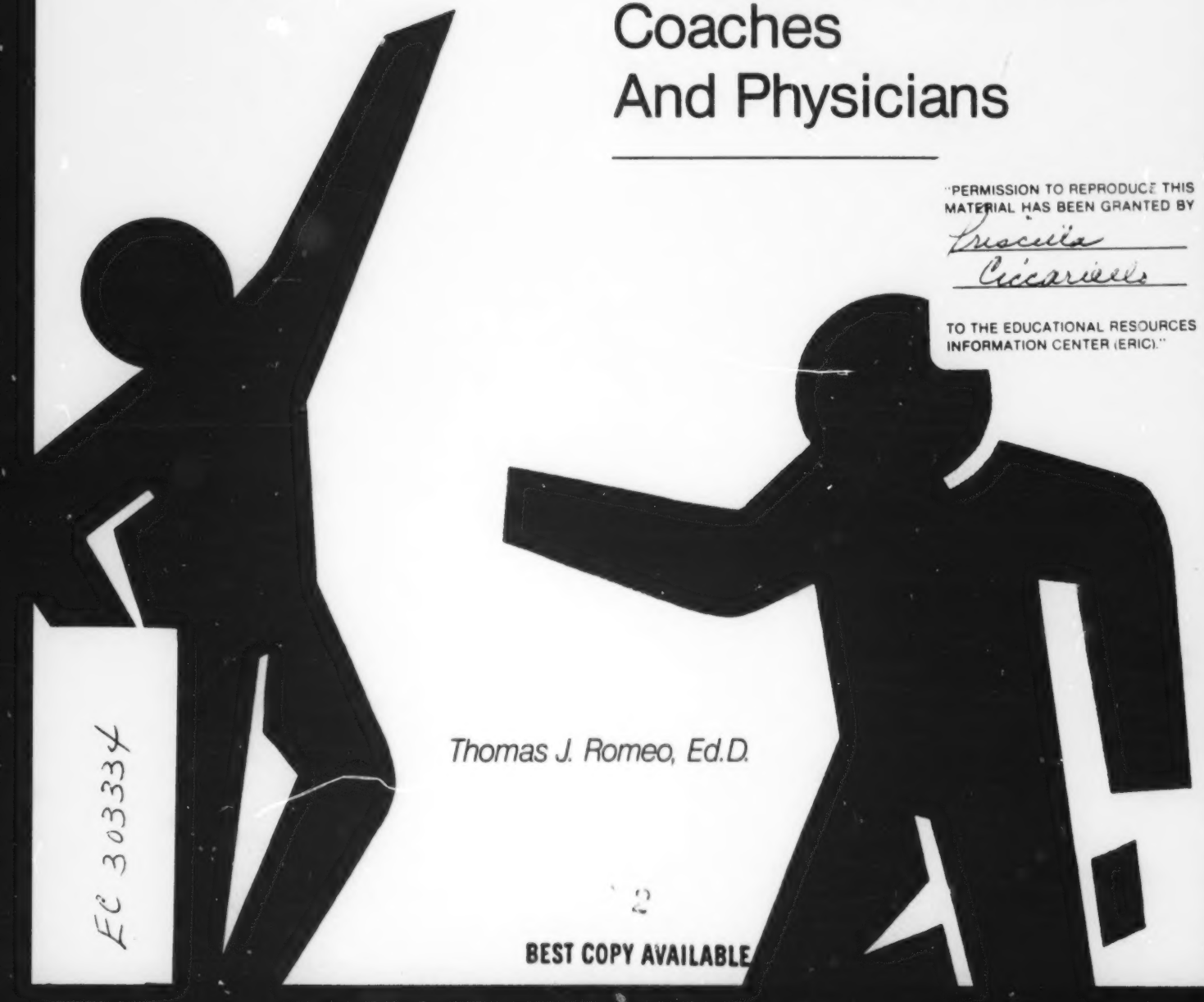

Thomas J. Romeo, Ed.D.

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Physical Manifestations of Marfan syndrome may vary dramatically from individual to individual. This manual can only be regarded as providing general guidelines, rather than specific recommendations, for any particular patient with Marfan syndrome. Specific recommendations can be provided to a patient only by a qualified physician. All patients with Marfan are urged strongly to contact a qualified physician and obtain that physician's approval before undertaking a physical activity program.

Neither the author nor the National Marfan Foundation assume responsibility for the physical activity program established/developed for a child with Marfan syndrome. This is solely the responsibility of the involved physician(s) and physical educator(s). The intent of this manual, and its contents, is to serve as a resource in providing information, suggestions, and guidelines, only.

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Physical Activity Guidelines for Physical Educators, Coaches and Physicians

Thomas J. Romeo, *Ed.D.*

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Preface

The Marfan syndrome is a heritable disorder of the connective tissue which occurs world-wide, in all races, and affects males and females equally. At the present time, it is estimated that approximately 40,000 individuals are afflicted with the syndrome in the United States, making it about as prevalent as cystic fibrosis, hemophilia, or muscular dystrophy. Because connective tissue is present throughout the body, multiple body systems are affected in varying degrees. The cause of the disorder is a defective protein, fibrillin, that is encoded by a gene on chromosome 15.

Major abnormalities may occur in the musculoskeletal, ocular, cardiovascular, and pulmonary systems of the body. The most serious effect of this genetic disorder is upon the cardiovascular system, where a widening of the aorta, if untreated or undiagnosed, may result in rupture and sudden death. Recently, much publicity has been focused upon the disorder because of the sudden death of Flo Hyman, a former star member of the United States Women's Olympic Volleyball Team.

At the present time, school-age Marfan children may be involved in, or excluded from, all aspects of physical education, including competitive athletics. Although it is desirable for most children with Marfan to participate in physical education, neither of these extremes is appropriate, unless placement is based upon sound medical and educational criteria.

This manual was developed for use by physical educators, coaches, and physicians seeking guidelines for prescribing physical activity for children with Marfan syndrome. The information contained in this manual will also be of interest to Marfan children and their parents, who are seeking information regarding appropriate physical activities for those diagnosed as having the disorder.

There presently exists a lack of knowledge on the part of the general public and medical and education professionals regarding the Marfan syndrome. Public Law 94-142, "The Education for All Handicapped Children Act," guarantees that an appropriate physical education program will be provided for all handicapped children between the ages of 3 and 21. This manual was written to provide guidance to professionals having responsibility for providing safe and effective physical activity programs for Marfan children in the public schools.

Two manuals published by the National Marfan Foundation (*The Marfan Syndrome* and *The Marfan Syndrome: A Booklet for Teenagers*) provide information related primarily to the physical and psychological ramifications of the disorder. It is hoped that this manual will provide those concerned with the physical education of Marfan children with the information needed to meet the mandates of P.L. 94-142, including physical activity guidelines and curriculum suggestions.

While the content of the manual is intended for use primarily by those providing physical activity for Marfan children in public school physical education classes, the information is also of value to coaches of competitive athletics. The tragic and sudden deaths of competitive athletes as a result of the Marfan syndrome underscores the importance of increased awareness of the disorder, its associated symptoms, and potential consequences.

The variability of the disorder, and differences in manifestations even within families, makes it extremely important to treat each child as an individual and with great care. The information provided in this document is, therefore, broad and general. The involved professionals must make selections for each Marfan child based solely upon that child's abilities, limitations, and needs. The importance of developing an Individualized Education Program (IEP) for each child, both for physical and psychosocial reasons, cannot be emphasized enough. The process requires cooperation, input, and ongoing assessment by all concerned.

It is expected that the information in the manual will clarify questions regarding physical activity for Marfan children. Therefore, the use of medical terminology has been minimized to allow for ease of reading and comprehension. It is hoped that in providing facts concerning the disorder, misinformation, and the fear and anxiety arising from such misinformation, will be minimized.

The content of this manual has been developed with the assistance of an advisory panel of educational, medical, and psychological experts. The manual includes information regarding P.L 94-142; physical characteristics associated with the disorder; and resultant physical needs. Also included is information regarding potential psychosocial implications; the establishment of educational goals to address these implications; as well as competencies and/or sensitivities needed by involved professionals. Suggested guidelines for physical activity, instructional methods and strategies, and approved curricular activities are included. In addition, contraindicated activities are identified and addressed as well as the implications for safety and effectiveness in physical activity programs. References and resources are also included.

The Marfan syndrome is one of many connective tissue disorders. The parameters, precautions, procedures and recommendations in this manual may be applicable to persons suffering from other connective tissue disorders to the extent that symptoms are similar to those associated with the Marfan syndrome. However, the guidelines in the manual should be applied to children with connective tissue disorders other than the Marfan syndrome only under the strict supervision of the treating physician. A program developed for a child with Marfan or any other connective tissue disorder should meet the unique individual needs, abilities and limitations of the involved child.

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Chapter 1

PUBLIC LAW 94-142: THE EDUCATION FOR ALL HANDICAPPED CHILDREN ACT

Goal

- To familiarize the reader with P.L. 94-142, "The Education for All Handicapped Children Act."

Objectives

- The reader will understand the terms "handicapped, special education, physical education, least restrictive environment, and Individualized Education Program (IEP)" as defined within the law.
- The reader will be informed of the guaranteed rights, due process, and legal actions available to parents/guardians of handicapped children.

Until P.L. 94-142 was created in 1975 for the specific purpose of guaranteeing a free and appropriate education for the millions of handicapped children in our country, provision of physical education for such children was rare to non-existent. A 1973 Bureau of Education for the Handicapped survey found that only a small percentage of all schools provided physical education for handicapped children, and of those that did, most provided inadequate programs.

A child identified as "handicapped" is guaranteed, under the provisions of the law, legal rights that must be addressed. The law, passed in 1975 by the federal government, is entitled, "P.L. 94-142, The Education for All Handicapped Children Act." The law defines the term, "handicapped," as including hearing impairments or deafness, visual impairments or blindness, deaf-blind impairments, speech impairments, emotional disturbance, mental retardation, orthopedic impairments, learning disabilities, other health related conditions, and multihandicapped.

The law, and its subsequent revisions, is very clear in intent and gives physical education special recognition as the only subject area mentioned by name in the definition of "Special Education." The definition of "Special Education" found in the law means specially designed instruction, at no cost to the parent, to meet the unique needs of a handicapped child, including classroom instruction, physical education instruction, home instruction, and instruction in hospitals and institutions.

This recognition demonstrates the importance that lawmakers place upon the discipline in the education of handicapped children. By comparison, recreation was assigned to the role of a "Related Service," and athletics and extra-curricular activities for the handicapped were placed under the title of, "Nonacademic Services."

"Physical Education" is defined, within the rules and regulations provided to implement the law, as the development of physical and motor fitness; fundamental motor skills and patterns; and skills in aquatics, dance, and individual and group games and sports (including intramurals and lifetime sports). Special physical education, adapted physical education, movement education, and motor development are included in the term "physical education."

While P.L. 94-142 requires that the education of handicapped children, ages 3 through 21, take place in the "least restrictive environment," that is, the most normal setting in which the child can succeed, its structure recognizes that the environment may indeed have necessary restrictions. When, as in the case of Marfan children, non-collision, non-isometric, low endurance activities are necessary, such limitations require the development of a specially designed program that may be outside the realm of "regular" or "mainstreamed" physical education.

Because the Marfan syndrome is a disorder affecting multiple body systems, children having the syndrome qualify under "handicapped" status due to impairments of individual body systems, or where more than one body system is affected, under the category of "other health impaired." While "labeling" of children continues to be a controversial topic for many parents, the services to be gained, and the potential benefits to the child, must be given careful consideration.

The parent of a child with Marfan syndrome should be familiar with P.L. 94-142 law as well as P.L. 93-112 because of the potential benefits these laws provide for a child. Section 504 of P.L. 93-112, entitled "The Rehabilitation Act of 1973," prohibits discrimination against handicapped individuals solely on the basis of their handicap, and requires that facilities be accessible to handicapped individuals under any program or activity which receives Federal financial aid.

Although physical education is guaranteed for a handicapped child under the law, it may be necessary to advocate for the level of quality which is appropriate for the child, and parents may need to become advocates for their child as well as other children with special needs. To that end, parents and professionals should familiarize themselves with these laws so that they may gauge whether their child's school is fulfilling the requirements of these laws by providing "free and appropriate" education and accessibility of facilities. In

some instances, it may be necessary for parents to join together to exert pressure upon school officials to bring about compliance with the provisions of P.L. 94-142. Individuals wishing to read the law in its entirety may request copies of the law from their local congressional representative.

A resource list of organizations that assist in identifying and addressing the legal rights and procedural guarantees of due process under P.L. 94-142 is included at the end of the manual. As a starting point, these are some important facts about the law:

- A free and appropriate education is guaranteed to every handicapped child under the law. Such education must be provided in an environment that is "least restrictive" according to the individual child's special needs and abilities.

- Children and parents are guaranteed procedural safeguards under the law. These include evaluation of the child by qualified individuals, and such evaluation may not take place unless permission is granted by the child's parents or guardian. In addition, time limits of 30 to 60 days are usually established regarding implementation of the special services once the process is initiated.

- An "Individualized Education Program" (IEP) must be developed for each handicapped child. Such a program will include a statement of present levels of performance; annual goals and short-term instructional objectives written in behavioral terms; where necessary, specific special education and related services to be provided; projected dates for initiation and duration of special services to be provided; and at least annual evaluation procedures to determine whether the short-term instructional objectives are being met. As an additional safeguard, parents must sign the plan to indicate approval of its contents.

- Related services, such as physical therapy, cannot be substituted for physical education instruction.

- In some states, a school district may be required to have a committee (Committee on Special Education) to rule on requests for special educational services, to oversee the educational placement and programs provided for children who are approved for special services, and to oversee the legal aspects of the law, including procedural safeguards.

- Under the law, the Commissioner of Education is expected to take action to insure that the mandated physical education services are provided to all handicapped children in schools.

If children with Marfan syndrome are not provided with physical education, or if the parent feels that the physical education program provided for the child is inappropriate, parents can play an important role. Within the law, the parent is recognized as a valuable participant in determining the education of the child and has mandated rights. Among these rights are educational assessments upon request, written notification prior to special testing, written notification prior to any change in educational placement, and a meeting with school representatives to discuss evaluation results and the provision of special support services.

Utilizing the results of the evaluation, the school must develop a written "Individualized Education Program," and meet with the parent within 30 days of the evaluation. If the parents do not agree with the results of the evaluation, or the I.E.P., they may refuse to sign the approval form. Following this, or any other disagreement, the parents are entitled to due process under the law. Some legal actions available to parents or guardians might include an appeal to local school representatives, a request for an independent evaluation, a request for a hearing before a neutral hearing officer, the filing of an administrative appeal, the filing of a complaint with the Federal Office for Civil Rights, or the filing of a lawsuit.

There is much more to the law than briefly indicated here. Parents of a child with Marfan syndrome should be encouraged to meet with the Director of Special Education in their local school district to discuss their expectations. Parents may wish to contact legislators. Parents may wish to become advocates for their child's special needs by joining local, state, and national advocacy groups for handicapped persons. Membership in the National Marfan Foundation is also an important consideration.

P.L. 94-142, and Section 504 of the Rehabilitation Act of 1973, are important laws affecting the lives of children with Marfan syndrome. Knowledge of the laws, and the ability to accurately recite or repeat important sections, is one of the most effective means of obtaining appropriate physical education programs for children who qualify for such consideration.

Chapter 2

PHYSICAL CHARACTERISTICS OF THE MARFAN SYNDROME

Goal

- To inform the reader as to the most common physical characteristics of the Marfan syndrome.

Objective

- The reader will be informed of the most common characteristics of the syndrome found in the musculoskeletal (muscles and bones), ocular (eyes), cardiovascular (heart and blood vessels), and pulmonary (lungs) systems.

Because of the diverse effects of the Marfan syndrome upon the various body systems, the physical characteristics associated with the disorder vary greatly with each individual. In fact, members of the same family who are affected by the disorder can show marked differences in physical characteristics (genetic heterogeneity).

The following physical characteristics, according to the body system affected, are those most commonly found in affected individuals:

Musculoskeletal System

Tall stature is commonly found in individuals affected by the Marfan syndrome, especially when it is accompanied by an elongation of the lower extremities. Elongation of the hands, fingers, legs, feet and toes (arachnodactyly) often occurs.

Deviations in the vertical line of the spine, including lateral deviations (scoliosis) or a combination of backward and lateral curvatures of the spine (kyphoscoliosis) are physical characteristics of the disorder. A sunken chest (pectus excavatum), protruding chest (pectus carinatum), or a combination of both, are also physical characteristics of the disorder.

Hyperextensible joints, especially in the knees (genu recurvatum), elbows and fingers, and flat feet (pes planus) are common characteristics of individuals affected by the syndrome. A decrease in muscle tone and muscle bulk, or both, are frequently associated with the disorder and contribute to the slender appearance of individuals with Marfan. Hernias, sometimes recurring in nature, are also among the most common physical Marfan characteristics.

Stretch marks (striae atrophicae) on the skin in the shoulder, chest, thigh, and/or lumbar areas, are also common, but do not impose any physical limitations. Finally, the mouth may present a high and/or narrow arched palate and crowding of teeth. Although stretch marks and crowded or crooked teeth do not normally present any significant physical problems, their cosmetic impact may be of significant psychosocial concern to the individual, especially to the adolescent.

Ocular System

The ocular system (eyes) is also affected by the Marfan syndrome. In fact one of the "tell-tale" signs denoting the potential presence of the Marfan syndrome is found in the ocular system in the form of dislocation of the lens of the eye (ectopia lentis). Near-sightedness (myopia) is another common characteristic associated with the ocular system.

Cardiovascular System

The effects of the syndrome upon the cardiovascular system pose the most serious threat to the affected individual because of their potentially life-threatening nature. These are most commonly manifested in the form of dilatation (widening) of the ascending aorta, mitral valve prolapse, and/or mitral regurgitation. A dangerously dilatated aorta can ultimately result in a tear in the wall of the aorta and sudden death. Dissection occurs when widening of the aorta causes a tear in the inner and middle layers of the aortic wall, resulting in the separation of these layers and the formation of a second channel through which blood is pumped. The force exerted by the blood pumping through this second channel may rupture the outer wall of the aorta.

Pulmonary System

The pulmonary system is most commonly affected by the increased potential for spontaneous collapse of part of the lungs (pneumothorax). Such an occurrence is usually manifested in sudden shortness of breath and chest pain, and, while not normally life-threatening, it may be extremely frightening.

Chapter 3

PHYSICAL ACTIVITY GUIDELINES

Goal

- To provide the reader with physical activity guidelines related specifically to the development of a physical activity program for Marfan children.

Objectives

- To provide the reader with guidelines related to physical needs, implications for safety and effectiveness, instructional methods and strategies, and sensitivities and competencies needed by the involved physical educator and physician.
- The reader will be informed of the physical education goals for children with Marfan syndrome based upon their identified physical needs.
- To provide the reader with a quick reference guide to programming for physical activity for children with Marfan syndrome.
- The reader will understand that each child is unique, and that it is the responsibility of the medical and educational professionals to establish appropriate parameters for exercise and to select specific activities based upon the child's unique abilities and needs.

Physical activity guidelines related specifically to the Marfan syndrome are crucial to the development of a physical education program that will be truly appropriate for a child with Marfan syndrome. Consideration of the physical needs of children with Marfan, implications for safety, implications for effectiveness, instructional methods and strategies, and sensitivities and competencies needed by the physical educator and physician, provide the bases for selection of curricular activities and parameters of exercise.

Such guidelines are intended to provide general information to those responsible for the development and implementation of physical activity programs for children with Marfan syndrome. In each case, however, it is the responsibility of the involved professionals to then establish appropriate parameters for exercise and to select specific activities based upon the child's unique abilities and needs.

Physical Needs of Children with Marfan and Physical Education Goals

In determining the most common physical characteristics associated with the Marfan syndrome, potentially related special physical needs arising from these characteristics were deduced. These needs, in turn, indicate potential goals for the adapted physical education program.

Physical characteristics found in one individual with Marfan may vary in degree and combination from those found in other affected individuals. The special needs of the particular individual, and the resultant goals of the physical education program will, therefore, vary accordingly.

Musculoskeletal System

Deviations in the vertical line of the spine, including scoliosis and kyphoscoliosis, require monitoring by an orthopedist and possibly bracing or surgery. Physical therapy may be required to strengthen muscles and ligaments supporting hyperextensible joints or to stretch muscles and ligaments where contractures exist. In addition, lack of muscle and bulk might have to be addressed through dietary means in an effort to maintain proper weight and strength. And finally, where the mouth and teeth are affected, orthodontia may be necessary to correct overcrowded and crooked teeth. Pectus deformities, such as a sunken or protruding chest, rarely require bracing or surgery, although they can impair normal functioning of the lungs. Most often, pectus deformities present problems of a cosmetic nature, rather than actually impairing functioning of the heart or lungs.

In response to these needs, the physical educator may have to provide exercises to strengthen and stretch muscles and ligaments, enhance body involvement, emphasize correct gross body mechanics, provide muscle toning activities, and improve poor posture mechanics.

Ocular System

The eyes are commonly affected by the disorder and require ongoing evaluation by an ophthalmologist, especially close monitoring of dislocated lenses. Corrective lenses are also required for near-sightedness (myopia), and may include eyeglasses or contact lenses. In some cases, bifocals may have to be worn over contact lenses. During physical activity, shatterproof eyeglasses must be worn or, in some cases, protective goggles.

The classroom teacher must be aware of visual limitations and seat the child accordingly. The physical educator must also take these limitations into consideration in order to place the child in an appropriate location during physical activity. The physical

educator must also remember that the child may have difficulty tracking objects and with visual perception. Therefore, consideration must be given to slowing down the speed of objects, providing progressive eye-limb coordination activities, and to the provision of visual perceptual-motor activities of a developmental nature.

The physical educator should be aware of the fact that prescription sun glasses may be necessary for outside activities (or those conducted in a very brightly lit indoor space) when the eyes must be kept partly dilated by medication. The physical educator should also be aware of the fact that vision with contact lenses may be profoundly different than vision without such lenses, and that contact lenses may fall out easily in Marfan children because of their flatter corneas.

In responding to the ocular needs of a child with Marfan syndrome, the physical educator may have to improve eye-hand and eye-foot coordination, provide opportunities to practice visual tracking of objects in motion (objects travelling near to far or far to near), and to assist in the development of other sequential perceptual motor skills (fine motor, gross motor, balance, spatial and body awareness).

Cardiovascular System

When a child is diagnosed with the Marfan syndrome, ongoing monitoring by a cardiologist familiar with the disorder is essential. Such monitoring usually occurs annually or semi-annually and may include echocardiography, electrocardiography, magnetic resonance imaging (MRI) and the prescription of medication. Although corrective cardiovascular surgery is a possibility for all Marfan patients, it is not usually necessary for children unless the aorta widens significantly or heart valves leak excessively and need repair or replacement.

Physical activity programs should be monitored by qualified physical educators, and individualized according to the child's special needs and medically approved parameters. Because of the potentially harmful effects upon the aorta of a child with Marfan syndrome, physical activity programs should be non-competitive in nature and should not include isometric activities. Activities such as swimming and bicycling should be of a progressive, submaximal aerobic nature within permissible tolerance levels established for the child by a physician.

The cardiovascular needs of children with Marfan require that the physical activity program achieve the following goals: a non-competitive activity environment, progressive submaximal aerobic activities, and the development of self-monitoring techniques (related to physical exertion limits) on the part of the child. It is also important to note that a

physical activity program designed for children with Marfan syndrome should include goals which address the learning of relaxation techniques, safety, comfort, and enjoyment.

Pulmonary System

Ongoing monitoring by a pulmonary specialist will insure proper medication where necessary, and avoidance of the potential for lung collapse (pneumothorax). Physical activity (such as swimming and walking) of appropriate length and load according to the child's individual needs and abilities should be provided. The activities should include breathing exercises and be selected on the basis of their potential ability to improve or maintain cardiopulmonary endurance.

Physical education goals related to the pulmonary needs of children with Marfan syndrome include the development of proper breathing techniques and the provision of activities which are appropriate in length and intensity load. The improvement and/or maintenance of the cardiopulmonary status of a child with Marfan are also physical education goals; these goals include teaching the child relaxation techniques to enhance safety, comfort, and enjoyment.

Implications for Safety

The physical activity program should be developed with the assistance of a multidisciplinary committee to insure that the program is safe (poses no undue risk of harm to the child). The multidisciplinary committee may include the treating physician (or treatment team, including the child's pediatrician, cardiologist, ophthalmologist, orthopedic surgeon, and pulmonary specialist), school doctor, school nurse, physical therapist, classroom teacher, and physical educator. The child and parents should participate in the decision-making process whenever possible.

Physical Activities Possibly Contraindicated for Children with Marfan

- Contact activities that might cause trauma to the chest, lungs, or heart, such as boxing, football, or wrestling. Trauma from such activities could cause the sudden collapse of a lung, or the dissection and/or rupture of a weakened or enlarged aorta.

- Activities that include a high risk of falling, such as gymnastics, horseback riding, or skating. The trauma from a fall could result in the collapse of a lung, or the dissection or rupture of a weakened or enlarged aorta.

- Isometric exercise activities. The increase in blood pressure and demand upon the heart and aorta could cause damage to the heart valves and/or dissection and rupturing of the aorta.

- Competitive sports (where aortic dilatation or mitral regurgitation exist). The demands of competitive sports upon a child with Marfan syndrome such conditions could result in dissection and/or rupturing of the aorta, or mitral valve leakage of fatal proportions.

- Demanding endurance activities, such as long distance or marathon running. The effects of such activities upon the lungs, heart, and aorta could result in collapse of a lung, further damage to the heart valves, and dissection or rupturing of the aorta.

- Diving and rapid decompression activities. The trauma from diving could cause damage to the lungs, heart, and/or aorta, and the effects of rapid decompression upon the heart and/or aorta could have potentially fatal results.

- Activities placing excessive strain upon joints and ligaments, such as gymnastics and wrestling. Such activities have the potential for causing serious injury to already weakened joints and ligaments.

- Activities placing excessive strain upon joints and ligaments, such as gymnastics and wrestling. Such activities have the potential for causing serious injury to already weakened joints and ligaments.

Suggested Safety Considerations According to Body Systems

Musculoskeletal System

- When bracing is necessary, the child's maneuverability, flexibility, speed, and endurance may be affected. Activities should be chosen accordingly.

- When a back or body brace is worn, the head and neck should be protected during physical activity by suitable padding.

- Activities that might cause dislocation of hyperextensible joints should be avoided.

- The physical education instructor should be informed by an orthopedist of the presence of restrictions upon range of motion and normal functioning.

- Children with flat feet must utilize properly fitting and supportive footwear during physical activity.

Ocular System

- Eye protection (goggles) should be utilized during physical activity when appropriate. However, it should be noted that there is presently no evidence supporting the contention that participation in physical activity leads to a progression in dislocation of the eye lens in individuals having the Marfan syndrome. The same is true of the suggestion that children with dislocated

eye lenses be excluded from participation in physical activity or sports because of danger of such progression. Prescription eyeglasses and frames that are safe for use during physical activity (shatter-proof, rounded edges) must be worn.

- The physical education instructor must be informed as to the child's vision problems and whether vision varies significantly with illumination or distance. The instructor must also be aware of any potential difficulty in following the flight of an object travelling far to near or near to far.

- The physical education instructor must be informed when contact lenses are worn because of the greater likelihood of their falling off of someone with the Marfan syndrome, especially during physical activity.

- Marfan children should be removed from physical activity and referred to the school nurse when they complain of vision problems, including flashing lights and sudden loss of vision.

Cardiovascular System

- The school must have written medical clearance by a cardiologist prior to permitting the child to participate in a physical activity program.

- Safeguards must be developed and maintained to prevent exertion at maximal levels. Submaximal levels of participation in aerobic activities are required.

- There must be avoidance of all trauma after valve replacement surgery because of anticoagulation medication and the risk of bleeding resulting from trauma. Careful monitoring of coagulation (prothrombin) time by medical personnel is required after valve replacement surgery. Any spontaneous bleeding or appearance of multiple bruises should be reported immediately to the school nurse.

- An audible valve click or noise is often heard when standing near a child with Marfan who has a prosthetic or artificial heart valve. This valve click is a normal phenomenon and not a cause for alarm.

- The physical educator must remain aware of the additional endurance limitations imposed upon a child who is wearing a brace.

- Periods of physical activity should be interspersed with periods of rest.

- Anaerobic activities which have the potential for large oxygen debt build-up must be avoided.

- Instruction in, and utilization of, relaxation techniques should be part of the physical activity program developed for children with Marfan.

- Appropriate water activities should be included (where facilities permit), especially those providing opportunities for relaxation and utilization of proper breathing techniques.

- Unnecessary circulatory restrictions during physical activity created by clothing or bracing must be eliminated.

- There must be an accurate understanding of the exercise tolerance limits of the involved child on the parts of both the physical education instructor and the child. As soon as children with Marfan syndrome are able, they should be instructed in self-monitoring techniques regarding the levels of intensity and duration which can be safely tolerated during physical activity.

- There should be gradual and progressive involvement in, and withdrawal from, physical activity through warm-up and cool-down phases of at least 5 to 7 minutes each.

- All school personnel, including the physical education instructor, must be informed if the child is taking medications. Information should include the purpose of the medication and its effect upon the body (beta-blockers effect the heart rate; blood thinner effects bruisability).

- Most children with Marfan syndrome receive beta-blockers to retard progressive widening of the aorta. Such medication will cause a slow pulse rate at rest and will blunt the normal rise in pulse that occurs with exercise. Therefore, achievement of an ideal pulse rate should not be expected in a child with Marfan syndrome who is exercising.

- Strategies must be developed to diffuse peer pressure on a child to participate in contraindicated physical activity. The physical educator should assist the child in developing a concept of self-competition, an understanding of the limitations imposed upon the body by the disorder, and the benefits of exercise and physical activity.

- School personnel involved in providing a physical activity program for children with Marfan should be proficient in CPR skills (especially as the protocols relate to the disorder), or have quick accessibility to those who do.

Pulmonary System

- There should be instructional emphasis upon correct breathing mechanics.

- An appropriate environment for physical activity, including consideration of hygiene, heat, and cold, must be provided in the school setting.

Implications for Effectiveness

Based upon the identified physical characteristics of a child with Marfan syndrome, a specially designed physical activity program can be developed to ameliorate the effects of the disorder on the affected body systems. In addition to being "safe," the program would have to be "effective, " that is, successfully meet the physical education needs of the child.

Listed below are suggestions for determining the effectiveness of an adapted physical education program in meeting the needs of a child with Marfan syndrome according to body systems affected by the disorder.

Musculoskeletal System

- Evaluation by the physician, physical therapist, and/or physical educator, of range of motion and joint laxity at the outset of the physical education program and re-evaluation at the conclusion of the school year.
- Assessment at the outset of the physical education program by the physician of joint pain and swelling and re-evaluation at the conclusion to determine whether the activities offered are appropriate or in need of revision.
- Monitoring by physician and/or school nurse of joint dislocations and injuries, as to frequency, site, and severity to determine whether the activities offered are appropriate or in need of revision.
- Periodic examination by the treating physician or pediatrician for occurrence and/or worsening of hernias during participation in a physical activity program to determine whether the activities offered are appropriate or in need of revision.
- Periodic utilization of objective strength testing during the physical activity program.
- Ongoing assessment by an orthopedist of posture through various means (visual, grid, x-ray), especially if a brace is utilized.
- Ongoing skinfold and height/weight measurements throughout the duration of the physical activity program.

Ocular System

- Information regarding the extent of visual impairment, including the presence or potential for dislocated lenses and retinal detachment, should be provided to the IEP planning committee and physical educator following examination by an ophthalmologist. Re-examination should take place at least annually to determine any changes and whether the physical activities offered are appropriate or in need of revision.
- Visual perceptual-motor evaluation should be completed at the outset of a physical activity program, including hand-eye and foot-eye coordination and at the end of each semester/year.

Cardiovascular and Pulmonary Systems

- A comprehensive cardiac examination should be completed prior to participation in a physical activity program, and repeated at least annually, or more frequently, as indicated by the cardiologist. This examination should include echocardiography (to assist in determining present levels of cardiac impairment, including changes in aortic dimension, the monitoring of cardiac valve functioning, and overall heart performance). Re-examination will determine if there have been any changes and whether the activities are appropriate or in need of revision.

- Ongoing monitoring of symptomatic levels of the child, including fatigue, chest and back pain, shortness of breath, arrythmias, and the occurrence of spontaneous bleeding or easy bruisability. These factors assist in determining changes in cardiovascular status and whether the activities are appropriate or in need of revision.

- Ongoing evaluation of imposed restrictions on physical activity to determine approriateness and/or the need for change.

- Observation of breathing mechanics of the child during rest, physical activity, and recovery to assist in determining whether the activities are appropriate or in need of revision.

- The development and monitoring of the child's ability to determine realistic personal exertion limits.

Instructional Methods and Strategies

The development of instructional methods and strategies, designed specifically to achieve the special psychosocial goals established for a child with Marfan syndrome, is an extremely important part of a comprehensive physical education program. Attention to these components will help maximize the potential for achieving goals determined to be the most positive and beneficial to the child.

Listed below are suggested instructional methods and strategies, categorized according to body systems affected by the disorder. These represent a suggested starting point; the quantity and quality of such methods and strategies are limited only by the instructor's imagination. Any approaches which assist in attaining a trusting, enjoyable, safe, and successful environment are acceptable.

General Methods

- Utilization of relaxing music as background for physical activity.
- Opportunities should be provided for the child to become familiar with the play area and surroundings prior to the initial start of a physical activity program.
- Wherever possible, aquatic activities should be utilized for their supporting and relaxing benefits.
- Rhythmic and dance activities should be employed to promote body awareness and control, and for the natural and special enjoyment they bring to movement.
- To minimize the potential for falling and thus increase confidence, require that only appropriate, properly fitting, supporting, and gripping footwear be worn by these children during physical activity.
- Include instruction in appropriate recreational and carry-over activities which the child with Marfan can engage in outside of the school setting and during adult life. This will enhance peer relationships and self-fulfillment.

Musculoskeletal System

- Select activities in which brace wearers can experience both enjoyment and success.
- Eliminate activities with potential for causing dislocation and thus decrease fear of injury and promote enjoyment and success.
- Select activities that are suitable for a child with Marfan syndrome who is suffering from joint arthritis and associated limitations in range of motion to promote success and eliminate fear of pain and injury.

Ocular System

- Adapt activities according to abilities, limitations, and needs, so that the child can overcome fears and be successful. Adaptations include:
    - using brightly colored objects
    - slowing speed/flight of object
    - using "soft" objects (foam, Nerf, rag)
    - using "sticking" aids (Velcro)
    - decreasing distance
- Utilize appropriate illumination to insure optimum visibility and to maximize the opportunity for success.
- Incorporate visual-motor activities to improve tracking skills and to promote feelings of success, accomplishment, and self-esteem.

- Require eye protection (goggles) during physical activity, where necessary, to reduce fears and timidity and to promote self-confidence.

- Select activities on the basis of the child's vision levels to insure success, decrease feelings of humiliation, stress, and poor body image, and to promote success and self-esteem.

Cardiovascular System

- Suggested adaptations related to this body system might include the following:

    - shortening/limiting length of play area
    - shortening time of activity
    - modifying implements (weight, height)
    - utilizing sitting or lying position in place of standing
    - utilizing required rest periods
    - utilizing greater numbers of participants
    - utilizing aquatics for support during activity
    - utilizing relaxation techniques
    - incorporating proper breathing techniques
    - eliminating unnecessarily "emotional," stressful, and/or, competitive activities

- Select activities requiring submaximal levels of exertion and include instruction in self-monitoring techniques regarding exercise intensity and tolerance levels to reduce fears and to increase children's feelings of control over their own destiny.

- Utilize warm-up and cool-down periods as required daily segments of physical activity participation for the Marfan child to provide for the necessary psychological and physical preparation for, and withdrawal from, physical activities.

- Require that the child's physician keep the physical education instructor informed regarding medications being taken, any changes in physical condition, and impending surgery in order for the instructor to be alert for signs of depression, lethargy, stress, social withdrawal, and/or rebellion.

- Select activities promoting the concept of self-competition to minimize the effects of peer pressure for the child to exceed physical exertion limits.

- Provide information to classmates regarding the Marfan syndrome so that they may better understand the effects of the disorder upon the body, provide support for the affected child, and refrain from teasing.

- Develop within the child a love and enjoyment of physical activity, as well as an understanding of its benefits in order to promote self-esteem and confidence, while eliminating unnecessary fears caused by ignorance.

Pulmonary System

- Provide instruction dealing with correct breathing mechanics to reduce stress and to promote enjoyment during physical activity.
- Provide instruction dealing with the selection of appropriate clothing for physical activity to insure comfort and to promote enjoyment.

Suggested Sensitivities and Competencies for Physical Educators

Ideally, the physical educator responsible for developing a physical activity program for a child with Marfan will be a specialist in "adapted physical education." However, this is not always the case, nor, does it have to be the case. A certified physical educator has the necessary training upon which to develop such a program, and upon which to become proficient in working with handicapped children.

The instructor must be willing to work with the child with Marfan, to deal with the related challenge, to follow the parameters permitted by medical professionals, and to locate and utilize any information or resources which may be of help. In addition, the physical educator must be prepared to use imaginative and innovative approaches to adapt activities, facilities, and equipment, and to continually work to improve existing professional skills and knowledge. Physical educators will need the encouragement, support, and assistance of school personnel, medical professionals, and parents, in order to provide the best possible physical education program for the child with Marfan.

At the outset, the adapted physical education specialist may have certain advantages over the regular physical educator by virtue of specialized training. In order to be effective, however, each will have to become familiar with the unique characteristics of the Marfan syndrome, as well as the unique characteristics, needs, and abilities of each affected child.

The following list represents suggested areas of competence and sensitivity which physical educators working with children with Marfan should possess or attempt to develop.

General Knowledge

- Willingness to learn as much as possible about Marfan syndrome and the affected child to minimize any potential for emotional or physical discomfort.
- Knowledge of the Marfan syndrome, and the ability to educate and sensitize parents, students, teachers, and administrators regarding the disorder.
- Knowledge of the physical characteristics of the Marfan syndrome and the resultant physical needs of children with Marfan syndrome.

- Knowledge of the potential psychosocial effects of the disorder upon affected children and their families, and a recognition of the pivotal role of parents.
- Knowledge of teaching techniques to improve the body image of children with Marfan syndrome.
- Knowledge of bracing techniques, their effect and limitations upon physical activity, and suitable activities for children wearing braces.
- Knowledge of physical activities and exercise which will improve the skills of children with Marfan syndrome.
- Knowledge of age appropriate activities in which these children can participate safely, successfully, and confidently, and the ability to apply limitations and adaptations accordingly.
- Knowledge of, and ability to evaluate, assessment devices used to determine current levels of ability.
- Knowledge of P.L. 94-142, and the ability to fulfill its mandates, including evaluation and development of the Individualized Education Program (IEP).
- Possess a general knowledge of normal growth and development, and of both handicapped and non-handicapped children in terms of psychomotor, cognitive, and affective abilities and needs.
- Knowledge of psychological development during childhood and adolescence, the ability to recognize emotional problems when they exist, and the ability to make appropriate referrals.
- Ability to develop trust and confidence on the part of the child with Marfan syndrome in the physical educator's ability to provide activities that are safe, as well as enjoyable.
- Knowledge of physical activity and exercise that will insure that the safety of the child will not be compromised.
- Ability to develop lessons that include information related to diet and nutrition that pertain specifically to the child.

Psychosocial Skills

- Sensitivity to the feelings experienced by children with the Marfan syndrome in school and physical education settings.
- The ability to recognize and monitor peer pressure thrust upon a child with Marfan syndrome within the school setting, and to positively offset such pressure when it might affect the safety or well-being of a child.
- The ability to minimize any potential for embarrassment through sensitivity and discretion.

- The ability to avoid patronizing children with Marfan syndrome by being completely honest and sincere in working with them.

- Sensitivity to individual differences in children regarding their ability to handle anxiety and stress.

- Understanding of the child's level of comprehension and the ability to communicate with the child accordingly. Often, because of the increased height, the child with Marfan may be treated as older than the child's actual age.

- A sensitivity to the child's psychosocial problems and resultant needs, as well as the ability to provide support when necessary.

Areas of Expertise

- The expertise to design activity progressions according to the abilities of the individual.

- The ability to participate and contribute as a member of the school's "Child Study Team," and/or the Committee on Special Education (CSE), regarding a child with Marfan syndrome.

- Demonstrate flexibility, high organizational skills, and the ability, especially in those cases where the adapted physical educator is an itinerant, to effectively communicate with other involved school personnel.

- The ability to develop a safe and effective physical education program for a child with Marfan syndrome, based upon that child's unique needs.

- The ability to select objectives and goals appropriate for the individual on the basis of present levels of performance and potential for improvement.

- The ability to monitor all aspects of the disorder as they relate to physical activity.

- Formal certification and the ability to administer First Aid and Cardio-Pulmonary Resuscitation (C.P.R.), specifically related to children with Marfan syndrome, since standard protocols may not be appropriate for a person with the disorder.

- The ability to educate the child regarding the importance and effects of physical activity upon the body, and to assist the child in developing a realistic self-concept of abilities and limitations.

- The ability to determine what a student can actually be expected to accomplish, so that neither too much, nor too little is expected, and the student is treated as normally as possible.

- The ability to observe, recognize, and communicate to the child with Marfan syndrome the positive aspects of personal growth and development.

- The ability to integrate individuals into class activities in accordance with needs, abilities, and limitations.
- Ability and willingness to serve as a positive adult role model for an affected child.
- A willingness to study and learn the potential effects of specific medications upon the student's performance and behavior.
- The ability to prescribe physical activity and exercise within the parameters permitted by physicians.
- Knowledge of relaxation techniques, and the ability to teach them to a child with Marfan syndrome.

Sensitivities and Competencies Related to Affected Body Systems

Musculoskeletal System

- A knowledge of the effects of exercise and movement upon joints, ligaments, and muscles, and the ability to select and design activities accordingly.
- The knowledge and ability to select physical activity to enhance proper posture and body alignment.
- The ability and willingness to communicate at regular intervals with the child's/adolescent's orthopedist and/or physical therapist to coordinate efforts to correct skeletal problems through appropriate physical activity.

Ocular System

- Ability to select, design and adapt physical activities which allow for visual deficiencies.
- Ability to select activities that minimize trauma to the eyes and head, and knowledge of those activities which require the use of protective eye devices by a child with Marfan syndrome.
- Ability to improve self-esteem and body image by promoting successful participation in physical activity despite a child's visual limitations and potentially related poor self-esteem and poor body image.
- Knowledge of visual-motor tests and motor development patterns in children with the Marfan syndrome.
- Knowledge of the effect of corrective eyeglasses or contact lenses upon the child's visual ability.
- Knowledge of terms used by ophthalmologists to express visual acuity, and an understanding of "best corrected visual acuity" versus "uncorrected visual acuity" concepts.

- Ability to select appropriate perceptual-motor activities for remedial or developmental purposes.

Cardiovascular System

- Understanding of the effect of beta-blockade medication upon the heart, and recognition of the fact that heart rate is not a true indicator of exertion level when such medication is in use.

- Knowledge of physical activities which, because of their vigorous nature, pose a risk of sudden damage to the aorta of a child with Marfan syndrome, and the need for avoidance or elimination of such activities.

- Understanding that a child with Marfan must not be pushed to exercise "through" symptoms when they occur, and the ability to recognize severe or changing symptoms requiring immediate referral to the child's physicians.

- Knowledge, understanding, and recognition of somnolence or lethargy, and associated decreases in the attention span of a child with Marfan syndrome.

- Knowledge of, and sensitivity to, the emotional aspects of the Marfan syndrome, including fear, anxiety, hopelessness, and helplessness, especially as related to the cardiac effects of the disorder.

- Knowledge of the effects of physical exertion upon the cardiovascular system in general, and the heart and aorta in particular.

- Ability to recognize when a child with Marfan syndrome is approaching his/her physical exertion limits.

- Ability and willingness to maintain close and ongoing communication with the child's cardiologist and other medical treatment team professionals.

- Understanding of the importance of providing rest periods during physical activity for the child with Marfan, and a willingness to utilize such breaks, whenever necessary.

Pulmonary System

- Knowledge regarding symptoms of sudden lung collapse (spontaneous pneumothorax), including sudden chest pain or shortness of breath, and emergency first-aid procedures to be utilized in such circumstances.

- Awareness that fear of recurrence of spontaneous lung collapse may be present in a child who has experienced a previous episode.

- Ability to provide an exercise environment in which an affected child can practice self-monitoring of activity tolerance and intensity.

- Understanding of the importance of providing a hygienically clean exercise environment, and one which minimizes extremes of hot and cold.

- Knowledge of correct breathing techniques, and the ability to teach these techniques to a child with Marfan syndrome.

- Knowledge of the aerobic demands of physical activity upon the pulmonary system of the child with Marfan syndrome and the ability to select appropriate activities accordingly.

- Ability to select appropriate physical activities in accordance with established medical parameters, when existing or potential pulmonary problems are identified.

Suggested Sensitivities and Competencies for Physicians

School or community-based general practitioners and/or pediatricians who serve children with Marfan syndrome must utilize all of their acquired skills to provide the best possible care for these special patients. Medical professionals must be willing to go that "extra step" if they are to meet the needs of these children. In addition to overseeing the health of the child throughout the year, they may be required to communicate frequently, both in written and verbal form, with the child's specialists and family members.

The child with Marfan syndrome spends much of life around doctors and hospitals and grows up relying more upon medical professionals than other children. The child faces the reality of mortality at a much younger age than most youngsters. Therefore, the child's primary care physician must deal with the psychosocial aspects of the Marfan syndrome.

As with the suggested competencies for physical educators working with these children, the following list represents suggested areas of competency and sensitivity which physicians should attempt to develop.

General Knowledge

- Possess a general knowledge of the Marfan syndrome, and the ability to educate the patient and family regarding the disorder, including its potential physical and psychological aspects.

- Knowledge of the genetic factors and effects associated with the Marfan syndrome so that other family members will be informed of their need to be evaluated by a specialist for the possible presence of the disorder.

- Knowledge of, and belief in, the importance of communication between the physician and physical educator regarding all testing and observations done on behalf of a child with the Marfan syndrome.

- Knowledge of, and belief in, the importance of the school staff and setting in providing a safe, successful, and trusting environment.

- Knowledge, ability, and willingness to provide the school staff with information addressing the importance of counseling and sensitizing of peers, faculty, and family regarding children with Marfan syndrome and their characteristics, abilities, and needs.

- A knowledge of childhood and adolescent psychosocial development, an awareness of the potential for emotional problems, and the ability to determine when and how to make appropriate referrals.

Psychosocial Skills

- Sensitivity to, and knowledge of, potential psychological feelings and psychosocial implications characteristic of handicapped children in general, and children with Marfan, in particular.

- Demonstrate a sensitivity to individual differences regarding stress and anxiety.

- Understanding the child's level of comprehension and communicating accordingly. Because the child with Marfan syndrome is taller than his or her peers, one must remember to treat the child age appropriately.

- Awareness of family dynamics and sensitivities, and an understanding of the pivotal role of parents when communicating.

Areas of Expertise

- The ability to recognize symptoms of the Marfan syndrome and to obtain appropriate confirmatory tests and opinions.

- Expertise in referring patients with Marfan syndrome to specialists or Marfan centers to insure that they receive appropriate consultations.

- Willingness to cooperate with Marfan specialists or a Marfan Team in order to better serve the child's needs and to become better informed concerning all aspects of the disorder.

- Ability to make recommendations to the physical educator regarding physical activities permitted, those contraindicated, implications of physical activity, and safe parameters for physical activity based upon a child's specific medical condition.

- The ability to be honest, forthright, and sincere when communicating findings and recommendations to avoid patronizing a child with Marfan.

- The ability to maintain an awareness of positive aspects of growth and development, and a belief in the need to address these when communicating with young patients with Marfan syndrome.

Sensitivities and Competencies Related to Affected Body Systems

Musculoskeletal System

- The family physician should possess knowledge of the spectrum of potential musculoskeletal problems associated with the syndrome.

- The orthopedist should possess knowledge of correct methods of physical diagnosis to determine skeletal abnormalities and changes not readily evident in the routine examination of a growing child or adolescent, including mild degrees of scoliosis and joint laxity.

- The orthopedist must demonstrate the ability to develop appropriate management techniques for orthopedic problems and to supervise the involved physical therapist(s).

- The orthopedist should demonstrate sensitivity to potential psycho-social implications of the Marfan syndrome as related to the effects of the disorder upon the musculoskeletal system.

- The orthopedist must possess knowledge of current bracing techniques and expertise in the appropriate and safe use of bracing during physical activity.

- The orthopedist must possess knowledge of current range-of-motion measurement techniques.

- The orthopedist must possess knowledge of the physiology of exercise, including musculoskeletal implications, range of motion parameters, and safety considerations.

- The orthopedist should demonstrate a willingness to communicate with the adapted physical educator regarding the development of a physical activity program based upon musculoskeletal implications, range of motion parameters, and safety considerations.

- Orthopedists must understand the risks associated with the use of anaesthesia during surgery performed to stabilize joints in a Marfan patient. In such situations, there must be a coordinated effort among the orthopedic surgeon, cardiologist, and anaesthesiologist.

- The orthopedist should possess knowledge of, and sensitivity to, the potential for anticipatory fear which a child may experience regarding impending surgery and the ability to provide support and reassurance during such periods.

- The orthopedist should possess the ability to prescribe appropriate and safe exercises to strengthen muscles surrounding lax joints.

- An orthopedist should be aware of the child's potential body image concerns and demonstrate sensitivity and discretion during physical examinations to minimize the potential for embarrassment.

- The orthopedist should demonstrate a willingness to refer a child with Marfan syndrome to a specialist, such as an Orthopedic Surgeon or Marfan Clinic, when it is necessary and appropriate.

- The orthopedist should possess the ability to develop appropriate techniques for the management of orthopedic problems and to recommend suitable exercises for use by a child with Marfan syndrome, in conjunction with the physical therapist, and/or physical educator. The physical therapist should work under orthopedic supervision.

Ocular System

- An ophthalmologist should possess knowledge of the spectrum of potential ocular problems associated with the syndrome, including dislocated lenses (ectopia lentis), near-sightedness (myopia), increased risk of retinal detachment, and relatively flat corneas.

- An ophthalmologist should demonstrate sensitivity to the potential for children with Marfan syndrome to feel uncomfortable wearing glasses or to deny their necessity as a result of feeling "different," and/or peer teasing. Sensitivity to the other related psychosocial implications of the disorder is helpful as well as a willingness to provide support and counseling.

- The family physician and/or optometrist should demonstrate a willingness and ability to refer the patient, when necessary, to an ophthalmologist who is expert in the diagnosis and treatment of ocular manifestations associated with the Marfan syndrome, and to maintain a close liaison when therapy or recommendations are necessary.

- The ophthalmologist should establish an ongoing schedule of eye examinations to monitor the potential effects of the disorder, and possess the necessary expertise and equipment to recognize ocular manifestations of the syndrome (such as early lens dislocation) often missed during routine eye examinations.

- The ophthalmologist should possess knowledge of the non-ocular features of the Marfan syndrome, since the eye specialist may be the first medical specialist to evaluate a patient who presents with dislocated lenses before the overall diagnosis of Marfan syndrome is made.

- The ophthalmologist should possess skill in aphakic and phakic refraction and utilization of good judgment in choosing which to use.

- The ophthalmologist should possess skill in indirect ophthalmoscopy for careful fundus examination.

- The ophthalmologist should possess the ability to explain why contact lenses may not be permitted, and honestly estimate when and if they will be permitted.

- The ophthalmologist should possess the ability to determine visual acuity, to provide appropriate care, to communicate limitations and needs, and to assist in the development of an appropriate physical activity program.

Cardiovascular System

- The family physician should possess knowledge of the spectrum of potential cardiovascular problems associated with the Marfan syndrome, including aortic dilatation, mitral valve prolapse, tricuspid valve prolapse, and mitral or aortic regurgitation.

- The family physician should demonstrate a willingness to refer a Marfan child to a cardiologist or Marfan center in order to obtain an appropriate cardiovascular diagnosis, treatment, and management plan.

- The family physician should possess a knowledge of the non-cardiovascular features of the Marfan syndrome, especially skeletal, so that a tall/thin child with unusually long arms and scoliosis who presents with mitral valve prolapse will be referred for further evaluation for suspected Marfan syndrome.

- The family dentist must prescribe endocarditis prophylaxis medication for Marfan patients with cardiovascular problems before and after treatment.

- The cardiologist should possess the ability to differentiate between Marfan patients needing marked physical activity restrictions and those who may be permitted wider participation parameters.

- The cardiologist should possess the ability to explain the rationale for restrictions to the child with Marfan, family, and family physician.

- The cardiologist should possess an understanding of the necessity and importance of on-going cardiac evaluation, at least annually, of providing school staff with significant information or changes, and maintaining open lines of communication.

- The cardiologist should possess the knowledge of the potential need for beta blockade, including expertise in prescribing beta blockade medication, its potential side effects, and its effect upon school and physical performance.

- The cardiologist should possess the ability to educate school personnel, especially the physical educator and nurse, regarding the prescription of beta blockade medication, its side effects, and impact upon physical activity.

- The cardiologist should possess the ability to prescribe a safe and realistic range of physical activity specific to the child's cardiovascular status, and to work closely with the involved physical educator to insure that such parameters are closely maintained.

- The cardiologist should possess the ability to diagnose changes in a Marfan patient's cardiovascular status, and to prescribe appropriate changes in medication and/or physical activity.

- The cardiologist should be aware that cardiac findings are progressive. Follow-up is needed and echocardiography is a critical element in such follow-up.

- The cardiologist should be able to determine the child's awareness of cardiac problems and to lend support in dealing with associated fears. Providing accurate information about medication and the necessity for adhering to medical directions helps in this regard.

Pulmonary System

- The family physician should be able to recognize that sudden chest pain and/or shortness of breath may be due to the collapse of a lung, and the need for referral to a hospital emergency room.

- The family physician should demonstrate a willingness to refer Marfan patients to an appropriate specialist(s) for baseline evaluation of pulmonary status and ongoing monitoring of findings.

- The pulmonary specialist should possess knowledge of the spectrum of pulmonary problems associated with the Marfan syndrome, including spontaneous lung collapse (pneumothorax).

- The pulmonary specialist should be sensitive to fear a child with Marfan may experience regarding spontaneous pneumothorax, especially if it has occurred previously; knowledge of preventative steps which the child can follow; and procedures for the child and/or school personnel to follow should the situation occur.

- The pulmonary specialist should understand the importance of emphasizing to school personnel the potentially harmful effects of extremes of hot and cold in the physical environment upon the pulmonary system of a patient with Marfan syndrome.

- The pulmonary specialist should be able to diagnose pulmonary problems associated with the Marfan syndrome and communicate information concerning the child's pulmonary status and management plan to the child, parents, physician, and school personnel.

- The pulmonary specialist should be aware of the non-pulmonary problems associated with the Marfan syndrome.

- The pulmonary specialist should be able, based on clinical findings, to prescribe appropriate and safe physical activity parameters to minimize spontaneous lung collapse or its recurrence.

- The pulmonary specialist should be aware of the fact that when lung collapse does occur in a patient with Marfan syndrome, the condition may not respond to the usual treatment modalities and may require surgical therapy.

- The pulmonary specialist should be able to develop an appropriate management plan for potential and/or existing pulmonary problems.

QUICK REFERENCE GUIDE
TO PROGRAMMING FOR PHYSICAL ACTIVITY FOR CHILDREN WITH MARFAN SYNDROME

PHYSICAL CHARACTERISTICS	PHYSICAL NEEDS	PSYCHOSOCIAL IMPLICATIONS	PSYCHOSOCIAL GOALS	INSTRUCTIONAL METHODS & STRATEGIES
MUSCULOSKELETAL SYSTEM:				
TALL STATURE	IMPROVED BODY MECHANICS SKILLS	SELF-CONSCIOUSNESS	REDUCE/ELIMINATE SELF-CONSCIOUSNESS	PROVIDE OPPORTUNITIES FOR SOCIAL INTERACTION DURING PHYSICAL ACTIVITY
	IMPROVED GROSS MOTOR SKILLS	PEER PRESSURE TO PLAY SPORTS	REDUCE/ELIMINATE PEER PRESSURE	EDUCATE/INFORM CLASSMATES ABOUT MARFAN SYNDROME EFFECTS
ELONGATION OF EXTREMITIES	IMPROVED FINE MOTOR SKILLS	POOR BODY IMAGE	IMPROVE BODY IMAGE	EMPLOY RHYTHMIC/DANCE ACTIVITIES TO PROMOTE POSITIVE BODY IMAGE/ CONTROL AND FOR NATURAL/SPECIAL ENJOYMENT THEY BRING TO MOVEMENT
	IMPROVED COORDINATION SKILLS	FEELING DIFFERENT/ UNATTRACTIVE	REDUCE/ELIMINATE FEELINGS OF DIFFERENCE/ UNATTRACTIVENESS	DISCUSS/EMPHASIZE DIFFERENCES AMONG ALL PEOPLE/HOW OTHERS WITH SIMILAR PROBLEMS "COPE"
KYPHOSCOLIOSIS	IMPROVED POSTURE/ HABITS	WITHDRAWING SOCIALLY	PROMOTE SOCIAL INVOLVEMENT	PROVIDE OPPORTUNITY FOR SOCIAL INTERACTION/ COOPERATION

PHYSICAL CHARACTERISTICS	IMPLICATIONS FOR SAFETY	IMPLICATIONS FOR EFFECTIVENESS	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICAL EDUCATOR	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICIAN
TALL STATURE	INSURE THAT BODY MECHANICS SKILL INSTRUCTION IS SEQUENTIAL AND APPROPRIATE	PRE/POST PARTICIPATION BODY MECHANICS SKILLS EVALUATION	KNOWLEDGE AND ABILITY TO SELECT PHYSICAL ACTIVITIES WHICH WILL IMPROVE BODY MECHANICS/ IMPROVE MOTOR SKILLS	KNOWLEDGE OF/SENSITIVITY TO POTENTIAL PHYSICAL AND PSYCHOSOCIAL IMPLICATIONS OF THE SYNDROME (UNUSUALLY TALL STATURE)
	INSURE THAT GROSS MOTOR SKILL INSTURCTION IS SEQUENTIAL AND APPROPRIATE	PRE/POST PARTICIPATION GROSS MOTOR SKILL EVALUATION		
ELONGATION OF EXTREMITIES	INSURE THAT FINE MOTOR SKILL INSTRUCTION IS SEQUENTIAL AND APPROPRIATE	PRE/POST PARTICIPATION FINE MOTOR SKILL EVALUATION	KNOWLEDGE AND ABILITY TO SELECT PHYSICAL ACTIVITIES WHICH WILL IMPROVE FINE MOTOR AND COORDINATION SKILLS	KNOWLEDGE OF THE SPECTRUM OF POTENTIAL MUSCULOSKELETAL EFFECTS OF THE DISORDER AND RESULTANT FINE/GROSS MOTOR PROBLEMS
KYPHOSCOLIOSIS		PRE/POST PARTICIPATION POSTURAL EVALUATION	KNOWLEDGE/ABILITY TO SELECT PHYSICAL ACTIVITY WHICH ENHANCES PROPER POSTURE/ BODY ALIGNMENT	ORTHOPEDIST MUST DEMONSTRATE ABILITY TO DEVELOP APPROPRIATE MANAGEMENT OF ORTHOPEDIC PROBLEMS

PHYSICAL CHARACTERISTICS	PHYSICAL NEEDS	PSYCHOSOCIAL IMPLICATIONS	PSYCHOSOCIAL GOALS	INSTRUCTIONAL METHODS & STRATEGIES
KYPHOSCOLIOSIS	BRACING	POOR SELF CONCEPT	IMPROVED SELF CONCEPT	PROVIDE OPPORTUNITIES FOR SUCCESS THROUGH PHYSICAL ACTIVITY AND REINFORCEMENT (MASTERY OF SKILLS; VERBAL REINFORCEMENT; MOTIVATION TO PERFORM)
SCOLIOSIS	BRACING	EMBARRASSMENT BECAUSE OF BRACE	REDUCE/ELIMINATE FEELINGS OF EMBARRASSMENT	PROVIDE PRIVACY FOR DRESSING/UNDRESSING AND SHOWERING
PECTUS DEFORMITIES	SURGERY	FEAR OF IMPENDING SURGERY	REDUCE/ELIMINATE FEARS	UTILIZE AQUATIC ACTIVITIES FOR THEIR SUPPORTING/RELAXING/ ENJOYMENT BENEFITS
	IMPROVED POSTURE HABITS	POOR BODY IMAGE	IMPROVE BODY IMAGE	PROVIDE ACTIVITIES THAT MAXIMIZE THE CHILD'S ABILITIES/ PHYSICAL CHARACTERISTICS AND INSURE ENJOYMENT AND SUCCESS

PHYSICAL CHARACTERISTICS	IMPLICATIONS FOR SAFETY	IMPLICATIONS FOR EFFECTIVENESS	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICAL EDUCATOR	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICIAN
KYPHOSCOLIOSIS	RANGE OF MOTION LIMITATIONS	RANGE OF MOTION MEASUREMENT	KNOWLEDGE OF BRACING AND RESULTANT PHYSICAL ACTIVITY LIMITATIONS	KNOWLEDGE OF BRACING TECHNIQUES; TAKING RANGE OF MOTION MEASUREMENTS
SCOLIOSIS	WHEN BRACE IS WORN, HEAD AND NECK SHOULD BE PROTECTED DURING PHYSICAL ACTIVITY BY SUITABLE PADDING	PRE/POST PARTICIPATION POSTURAL EVALUATION	KNOWLEDGE/WILLINGNESS TO COMMUNICATE WITH CHILD'S ORTHOPEDIST/PHYSICAL THERAPIST TO COORDINATE EFFORTS TO CORRECT MUSCULOSKELETAL PROBLEMS THROUGH APPROPRIATE PHYISCAL ACTIVITY	A KNOWLEDGE OF CURRENT BRACING TECHNIQUES, AND EXPERTISE IN THE APPROPRIATE AND SAFE UTILIZATION OF BRACING DURING PHYSICAL ACTIVITY
PECTUS DEFORMITIES	FURTHER RESTRICTION OF PHYSICAL ACTIVITY IS REQUIRED FOLLOWING PECTUS SURGERY	EVALUATION BY SURGEON REGARDING RETURN TO PHYSICAL ACTIVITY AND MONITORING BY PHYSICAL EDUCATOR/NURSE UPON RETURN	SENSITIVITY TO/AWARENESS OF POTENTIAL PSYCHOSOCIAL IMPLICATIONS ASSOCIATED WITH PECTUS DEFORMITIES, ESPECIALLY AS RELATED TO ADOLESCENTS	SENSITIVITY TO/AWARENESS OF POTENTIAL PSYCHOSOCIAL IMPLICATIONS ASSOCIATED WITH PECTUS DEFORMITIES ESPECIALLY AS RELATED TO ADOLESCENTS

PHYSICAL CHARACTERISTICS	PHYSICAL NEEDS	PSYCHOSOCIAL IMPLICATIONS	PSYCHOSOCIAL GOALS	INSTRUCTIONAL METHODS & STRATEGIES
HERNIAS	TRUSSING	EMBARRASSMENT BECAUSE OF TRUSS	REDUCE EMBARRASSMENT REGARDING TRUSS	PROVIDE PRIVACY FOR DRESSING/UNDRESSING/SHOWERING
	IMPROVED LIFTING SKILLS/MECHANICS	FEAR OF RECURRENT PAIN	REDUCE FEAR AND PROMOTE CONFIDENCE	PROVIDE OPPORTUNITY FOR CHILD TO ASSIST IN SELECTION OF ACTIVITIES IN WHICH ENJOYMENT AND SUCCESS ARE OUTCOMES
FLAT FEET	SUPPORT ASSISTANCE (ORTHOTICS/FOOTWEAR)	LACKING SELF-CONFIDENCE	BUILD SELF-CONFIDENCE	PROVIDE ACTIVITIES IN WHICH CHILD WILL BE SUCCESSFUL
HYPEREXTENSIBLE JOINTS	PHYSICAL THERAPY	FEELING OF LOW SELF-ESTEEM	IMPROVE SELF-ESTEEM	DISCUSS DIFFERENCES WHICH EXIST AMONG ALL PEOPLE AND PROVIDE EXAMPLES/ROLE MODELS OF PEOPLE WHO HAVE SUCCEEDED

PHYSICAL CHARACTERISTICS	IMPLICATIONS FOR SAFETY	IMPLICATIONS FOR EFFECTIVENESS	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICAL EDUCATOR	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICIAN
HERNIAS	PROVISION OF ACTIVITIES WHICH MINIMIZE LIFTING/ CLIMBING	EVALUATION FOR HERNIA BY PHYSICIAN TO DETERMINE OCCURRENCE/RECURRENCE DUE TO PHYSICAL ACTIVITY	SENSITIVITY TO POTENTIAL PSYCHOSOCIAL IMPLICATIONS RELATED TO WEARING OF TRUSS/ SURGERY	A KNOWLEDGE OF SYMPTOMS/TREATMENT REGARDING HERNIAS AND RELATED RESTRICTIONS REGARDING PHYSICAL ACTIVITY
FLAT FEET	MONITORING BY PHYSICAL EDUCATOR TO INSURE THAT APPROPRIATE FOOTWEAR IS WORN	MONITORING BY PHYSICIAN OF FOOTWEAR/ ORTHOTICS AND CONDITION OF FEET/ASSOCIATED PAIN	KNOWLEDGE OF EFFECT OF FLAT FEET UPON BODY ALIGNMENT/ MECHANICS/MOVEMENT	KNOWLEDGE OF SYMPTOMS/ TREATMENT REGARDING FLAT FEET AND RESULTANT IMPACT UPON BODY MECHANICS/ MOVEMENT
HYPEREXTENSIBLE JOINTS	ACTIVITIES THAT MIGHT CAUSE DISLOCATION OF HYPEREXTENSIBLE JOINTS SHOULD BE AVOIDED	EVALUATION BY THE PHYSICIAN, PHYSICAL EDUCATOR, AND/OR NURSE OF RANGE OF MOTION/JOINT LAXITY/ DISLOCATIONS AT OUTSET OF THE PHYSICAL ACTIVITY PROGRAM AND RE-EVALUATION AT THE END OF THE SCHOOL YEAR	KNOWLEDGE OF THE EFFECTS OF EXERCISE AND MOVEMENT UPON JOINTS, LIGAMENTS, AND MUSCLES, AND THE ABILITY TO SELECT AND DESIGN ACTIVITIES ACCORDINGLY	POSSESS KNOWLEDGE OF THE PHYSIOLOGY OF EXERCISE, INCLUDING MUSCULOSKELETAL IMPLICATIONS, RANGE OF MOTION PARAMETERS AND SAFETY CONSIDERATIONS

PHYSICAL CHARACTERISTICS	PHYSICAL NEEDS	PSYCHOSOCIAL IMPLICATIONS	PSYCHOSOCIAL GOALS	INSTRUCTIONAL METHODS & STRATEGIES
HYPEREXTENSIBLE JOINTS	MUSCLE/LIGAMENT STRENGTHENING	FEELING SELF-CONSCIOUS	REDUCE/ELIMINATE SELF-CONSCIOUSNESS	ELIMINATE ACTIVITIES WITH A GREATER POTENTIAL FOR CAUSING DISLOCATIONS, DECREASING FEAR/PROMOTING CONFIDENCE
LACK OF MUSCLE TONE/ BULK	INCREASED MUSCLE-TONE	FEELING LESS MASCULINE/ FEMININE	PROMOTE FEELINGS OF MASCULINITY/FEMININITY	PROVIDE ACTIVITIES (DANCE) THAT EMPHASIZE MALE/FEMALE ROLES
	DIETARY CONTROLS	FEELING "DIFFERENT"	REDUCE/ELIMINATE FEELINGS OF DIFFERENCE	EMPHASIZE ABILITIES/ QUALITIES OF THE CHILD THROUGH SELECTED ACTIVITIES AND LEARNING EXEPERIENCES
CONTRACTURES	MUSCLE/LIGAMENT STRETCHING	FEELING HUMILIATED	REDUCE/ELIMINATE FEELINGS OF HUMILIATION	SELECT ACTIVITIES THAT ARE SUITABLE FOR THE CHILD SUFFERING FROM JOINT ARTHRITIS AND ASSOCIATED LIMITATIONS IN RANGE OF MOTION TO PROMOTE SUCCESS AND ELIMINATE FEELINGS OF HUMILIATION

PHYSICAL CHARACTERISTICS	IMPLICATIONS FOR SAFETY	IMPLICATIONS FOR EFFECTIVENESS	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICAL EDUCATOR	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICIAN
LACK OF MUSCLE-TONE/ BULK	PROVIDE ACTIVITIES APPROPRIATE TO CHILD'S STRENGTH/ENDURANCE	ONGOING ASSESSMENT OF ACTIVITIES BY PHYSICAL EDUCATOR AS TO APPROPRIATENESS/ UTILIZATION OF OBJECTIVE AND APPROPRIATE STRENGTH/WEIGHT TESTING	KNOWLEDGE OF OBJECTIVE AND APPROPRIATE STRENGTH TESTING DURING THE PHYSICAL ACTIVITY PROGRAM	KNOWLEDGE OF SKINFOLD, HEIGHT, AND WEIGHT EVALUATIONS TO MONITOR CHILD'S MUSCLE TONE/BULK THROUGHOUT THE DURATION OF THE PHYSICAL ACTIVITY PROGRAM
CONTRACTURES	PROVIDE ACTIVITIES APPROPRIATE TO CHILD'S RANGE OF MOTION PARAMETERS IN JOINTS AFFECTED BY CONDITION(S)	MONITORING OF CONTRACTURES BY PHYSICAL EDUCATOR, NURSE, AND/OR PHYSICIAN FOR CHANGES IN EXISTING CONTRACTURES	KNOWLEDGE OF RANGE OF MOTION EVALUATION DEVICES TO MONITOR CONTRACTURES REGARDING CHANGES IN CONDITION(S)	EXPERTISE IN MONITORING OF CONTRACTURES TO DETERMINE IF CHANGES IN TREATMENT ARE NECESSARY, TO COORDINATE TREATMENT WITH THE PHYSICAL THERAPIST, AND TO RECOMMEND SURGICAL INTERVENTION WHEN APPROPRIATE

PHYSICAL CHARACTERISTICS	PHYSICAL NEEDS	PSYCHOSOCIAL IMPLICATIONS	PSYCHOSOCIAL GOALS	INSTRUCTIONAL METHODS & STRATEGIES
CONTRACTURES	PHYSICAL THERAPY	FEELING "DIFFERENT"	REDUCE/ELIMINATE FEELINGS OF DIFFERENCE	EMPHASIZE ABILITIES/ QUALITIES OF THE CHILD THROUGH SELECTED ACTIVITIES AND LEARNING EXPERIENCES
HIGH/NARROW ARCHED PALATE/CROWDED TEETH	ORTHODONTIA	FEELING EMBARRASSED BECAUSE OF ORTHODONTIA	ELIMINATE FEELINGS OF EMBARRASSMENT	UTILIZE INFORMAL DISCUSSIONS TO POINT OUT THE LARGE NUMBER OF CHILDREN WHO WEAR BRACES/CONGRATULATE OTHER CHILDREN/PEERS WHO HAVE OR HAVE HAD BRACES
STRETCH MARKS	NONE (COSMETIC IN NATURE)	DRESSING/UNDRESSING INHIBITIONS	REDUCE/ELIMINATE INHIBITIONS	PROVIDE FOR PRIVACY IN DRESSING/UNDRESSING/ SHOWERING
OCULAR SYSTEM:				
DISLOCATED EYE LENSES	OPHTHALMOLOGICAL MONITORING	FEELING SELF-CONSCIOUS	REDUCE/ELIMINATE FEELINGS OF SELF-CONSCIOUSNESS/ FEELINGS OF BEING HANDICAPPED	PLAN ACTIVITIES BASED UPON THE STUDENT'S ABILITIES AND WHICH PROMOTE THE CHILD'S INDEPENDENCE/DECISION MAKING
	SURGERY	FEELING "HANDICAPPED"		

PHYSICAL CHARACTERISTICS	IMPLICATIONS FOR SAFETY	IMPLICATIONS FOR EFFECTIVENESS	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICAL EDUCATOR	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICIAN
HIGH/NARROW ARCHED PALATE/CROWDED TEETH	UTILIZATION (WHEN NECESSARY) OF MOUTHPIECE TO PROTECT TEETH/MOUTH DURING PHYSICAL ACTIVITY	EVALUATION OF THE MOUTH/TOOTH AREA BY ORTHODONTIST (PRE/POST PHYSICAL EDUCATION PARTICIPATION) TO DETERMINE ORAL STATUS	SENSITIVITY TO THE PSYCHOSOCIAL IMPACT UPON A CHILD WEARING BRACES/PROTECTIVE MOUTHPIECE	EXPERTISE (ORTHODONTIST) REGARDING THE USE OF BRACES TO CORRECT DENTAL MALOCCLUSION
STRETCH MARKS	NO SPECIAL SAFETY CONSIDERATIONS	NO SPECIAL EFFECTIVENESS CONSIDERATIONS	SENSITIVITY TO THE PSYCHOSOCIAL EFFECTS OF STRETCH MARKS, ESPECIALLY ON ADOLESCENTS	SENSITIVITY TO THE PSYCHOSOCIAL EFFECTS OF STRETCH MARKS, ESPECIALLY ON ADOLESCENTS
OCULAR SYSTEM:				
DISLOCATED EYE LENSES	COMPLAINTS OF VISION PROBLEMS OF ANY KIND (FLASHING LIGHTS/SUDDEN LOSS OF VISION) MUST BE TAKEN SERIOUSLY BY THE PHYSICAL EDUCATOR; CHILD MUST BE REMOVED FROM PHYSICAL ACTIVITY AND REFERRED TO NURSE	PRE/POST PHYSICAL ACTIVITY PARTICIPATION EVALUATION OF VISUAL ACUITY	THE ABILITY TO SELECT ACTIVITIES THAT MINIMIZE THE POTENTIAL FOR TRAUMA TO THE EYES AND HEAD AND REQUIRE UTILIZATION OF PROTECTIVE EYE DEVICES WHEN NECESSARY	(OPHTHALMOLOGIST) DEMONSTRATE THE ABILITY TO DETERMINE VISUAL ACUITY, TO PROVIDE APPROPRIATE CARE, TO COMMUNICATE LIMITATIONS AND NEEDS, AND TO ASSIST IN THE DEVELOPMENT OF AN APPROPRIATE PHYSICAL ACTIVITY PROGRAM

PHYSICAL CHARACTERISTICS	PHYSICAL NEEDS	PSYCHOSOCIAL IMPLICATIONS	PSYCHOSOCIAL GOALS	INSTRUCTIONAL METHODS & STRATEGIES
NEAR-SIGHTEDNESS	CORRECTIVE LENSES (EYEGLASSES/CONTACTS)	PEER TEASING	REDUCE/ELIMINATE PEER TEASING	EDUCATE/INFORM CLASSMATES ABOUT MARFAN SYNDROME TO INCREASE UNDERSTANDING/SENSITIVITY/ACCEPTANCE
	IMPROVED PERCEPTUAL MOTOR SKILLS	DIFFICULTY FUNCTIONING	REDUCE EMBARRASSMENT AND SELF- CONSCIOUSNESS WHEN FUNCTIONING IN CLASS	UTILIZE VISUAL-MOTOR ACTIVITIES TO IMPROVE EYE TRACKING SKILLS AND TO PROMOTE FEELINGS OF SUCCESS/ ACCOMPLISHMENT AND SELF-ESTEEM
CARDIOVASCULAR SYSTEM:				
DILATATION OF THE AORTA	CARDIAC MONITORING OF AORTIC ROOT DIMENSION	FEELING DEPRESSED	REDUCE/ELIMINATE FEELINGS OF DEPRESSION	PROVIDE ACTIVITIES WHICH ARE FUN AND WHICH PROMOTE POSITIVE PEER INTERACTION
	BETA BLOCKADE MEDICATION	REFUSING TO TAKE MEDICATION	PROMOTE FEELINGS OF COOPERATION	PROVIDE LEARNING ENVIRONMENT THAT ENCOURAGES COMMUNICATION/SUPPORT/TEAM WORK/EMPHASIZES SELF-WORTH

PHYSICAL CHARACTERISTICS	IMPLICATIONS FOR SAFETY	IMPLICATIONS FOR EFFECTIVENESS	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICAL EDUCATOR	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICIAN
NEAR-SIGHTEDNESS	PRESCRIPTION EYE-GLASSES AND FRAMES THAT ARE SAFE FOR USE DURING PHYSICAL ACTIVITY (SHATTER-PROOF/ ROUNDED EDGES) MUST BE WORN	PRE/POST PHYSICAL ACTIVITY PARTICIPATION EVALUATION OF VISUAL ACUITY	THE ABILITY TO SELECT/ DESIGN PHYSICAL ACTIVITIES WHICH CONSIDER VISUAL NEEDS ANDTHE ABILITY TO USE DEVELOPMENTAL AND/ OR REMEDIAL APPRAOCHES	SENSITIVITY TO THE POTENTIAL FOR MARFAN CHILDREN TO FEEL UNCOMFORTABLE WEARING EYEGLASSES OR TO DENY THEIR NECESSITY AS A RESULT OF FEELING "DIFFERENT" AND/OR PEER TEASING, AS WELL AS OTHER RELATED PSYCHOSOCIAL IMPLICATIONS
CARDIOVASCULAR SYSTEM:				
DILATATION OF THE AORTA	SCHOOLS MUST HAVE WRITTEN MEDICAL CLEARANCE BY A CARDIOLOGIST PRIOR TO PERMITTING A CHILD TO PARTICIPATE IN A PHYSICAL ACTIVITY PROGRAM	A COMPREHENSIVE CARDIAC EXAMINATION SHOULD BE COMPLETED PRIOR TO PARTICIPATION IN A PHYSICAL ACTIVITY PROGRAM AND REPEATED AT LEAST ANNUALLY	A KNOWLEDGE OF PHYSICAL ACTIVITIES WHICH, BECAUSE OF THEIR VIGOROUS NATURE, POSE A RISK OF DAMAGE TO THE AORTA AND THE NEED FOR ADAPTATION OR CONTRAINDICATION	(CARDIOLOGIST) KNOWLEDGE OF THE POTENTIAL NEED FOR BETA BLOCKADE MEDICATION AND PHYSICAL ACTIVITY RESTRICTION IN THE TREATMENT OF MARFAN PATIENTS, INCLUDING EXPERTISE IN PRESCRIBING BETA BLOCKADE MEDICATION, POTENTIAL SIDE EFFECTS UPON SCHOOL AND PHYSICAL PERFORMANCE
	SCHOOL PERSONNEL MUST BE INFORMED WHEN A CHILD IS TAKING CARDIOVASCULAR MEDICATIONS (ESPECIALLY BETA-BLOCKADE MEDICATION)	ONGOING COMMUNICATION AMONG ALL INVOLVED IN THE CHILD'S PHYSICAL ACTIVITY PROGRAM		

PHYSICAL CHARACTERISTICS	PHYSICAL NEEDS	PSYCHOSOCIAL IMPLICATIONS	PSYCHOSOCIAL GOALS	INSTRUCTIONAL METHODS & STRATEGIES
MITRAL VALVE PROLAPSE	STRESS REDUCTION BY PARENTS	FEELING OVER-PROTECTED	PROMOTE FEELINGS OF INDEPENDENCE/RESPONSIBILITY	ALLOW STUDENT TO PARTICIPATE IN PLANNING OF PROGRAM/ ACTIVITIES AND ASSIGN DUITES/LEADERSHIP RESPONSIBILITIES
	PROFICIENCY IN RELAXATION SKILLS	FEELING ANGRY/ACTING OUT	REDUCE/ELIMINATE ANGER/ACTING OUT	PROVIDE OPPORTUNITIES FOR "VENTING" OF FEELINGS/FRUSTRATIONS AND PHYSICAL ACTIVITY WHICH SERVES AS AN OUTLET FOR ENERGY RELEASE
PULMONARY SYSTEM:				
SPONTANEOUS PNEUMOTHORAX	PULMONARY MONITORING	FEAR OF OCCURRENCE/ RECURRENCE OF PNEUMOTHORAX	ELIMINATE/MINIMIZE FEARS	PROVIDE OPPORTUNITIES FOR VERBALIZATION OF FEARS AND INFOMATION RELATED TO FIRST AID PLAN IN THE EVENT SUCH AN INCIDENT OCCURS/ RECURS PROVIDE ACTIVITIES/ OPPORTUNITIES WHICH PROMOTE LEADERSHIP/ INITIATIVE/FORCEFULNESS

PHYSICAL CHARACTERISTICS	IMPLICATIONS FOR SAFETY	IMPLICATIONS FOR EFFECTIVENESS	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICAL EDUCATOR	SENSITIVITIES/ COMPETENCIES NEEDED BY PHYSICIAN
MITRAL VALVE PROLAPSE	PERIODS OF PHYSICAL ACTIVITY MUST BE INTERSPERSED WITH PERIODS OF REST	MONITORING OF SYMPTOMATIC LEVELS OF THE CHILD, INCLUDING FATIGUE, CHEST AND BACK PAIN, SHORTNESS OF BREATH, ARRYTHMIAS, AND THE OCCURRENCE OF SPONTANEOUS BLEEDING OR EASY BRUISABILITY	A KNOWLDGE OF THE EFFECTS OF PHYSICAL EXERTION UPON THE CARDIOVASCULAR SYSTEM GENERALLY, AND THE HEART AND THE AORTA, SPECIFICALLY	(CARDIOLOGIST) THE ABILITY TO PRESCRIBE A SAFE AND REALISTIC RANGE OF PHYSICAL ACTIVITY SPECIFIC TO THE CHILD'S CARDIOVASCULAR STATUS AND TO WORK WITH THE PHYSICAL EDUCATOR TO INSURE THAT SUCH PARAMETERS ARE MAINTAINED
PULMONARY SYSTEM:				
SPONTANEOUS PNEUMOTHORAX	AN APPROPRIATE ENVIRONMENT MUST BE PROVIDED FOR PHYSICAL ACTIVITY INCLUDING CONSIDERATION OF HYGIENE, HEAT, AND COLD CONTRAINDICATION OF ALL DIVING AND RAPID DECOMPRESSION ACTIVITIES	ONGOING MONITORING OF THE CHILD'S BREATHING MECHANICS DURING REST AND ACTIVITY ONGOING REEVALUATION OF IMPOSED RESTRICTIONS ON PHYSICAL ACTIVITY TO DETERMINE APPROPRIATENESS AND/ OR NEED FOR REVISION	KNOWLEDGE REGARDING SYMPTOMS OF SUDDEN LUNG COLLAPSE (PNEUMOTHORAX), INCLUDING CHEST PAIN OR SHORTNESS OF BREATH, AND EMERGENCY FIRST-AID PROCEDURES TO BE FOLLOWED IN SUCH CIRCUMSTANCES	KNOWLEDGE OF THE SPECTRUM OF PULMONARY PROBLEMS ASSOCIATED WITH THE MARFAN SYNDROME, INCLUDING SPONTANEOUS LUNG COLLAPSE

Chapter 4

PSYCHOSOCIAL IMPLICATIONS AND GOALS IN ADAPTED PHYSICAL EDUCATION FOR CHILDREN WITH MARFAN

Goal

- To provide the reader with information regarding the potential psychosocial implications of the physical effects of the Marfan syndrome and the related physical education goals

Objectives

- The reader will be informed of potential psychosocial implications resulting from the physical effects of the syndrome.
- The reader will be informed of the psychosocial goals of an adapted physical education program for children with Marfan syndrome.

Psychosocial Implications

Anything that causes someone to be "different" from others is a potential source of psychosocial (psychological and social) difficulty. The potential psychosocial implications of the various physical effects of the Marfan syndrome will vary with each individual.

Within the home, the attitude of the child's parents, the environment in which the child is raised, and the age of the child when the disorder is diagnosed, are critical factors influencing the psychosocial effects of a disorder. Within the school setting, the teacher's awareness of the disorder, attitude toward the child, and ability to deal with the child and the child's classmates are equally important. The psychosocial implications are directly related to, and affected by, these factors.

Potential psychosocial implications for children having the Marfan syndrome include:

- feelings of self-consciousness
- feeling "different"
- feeling unattractive
- feeling humiliated
- feeling depressed
- feeling less masculine or feminine
- feeling embarrassed
- feeling "handicapped"
- feeling isolated

- feeling hopeless
- feeling timid
- withdrawing socially
- feeling stressed
- feelings of low-self-esteem
- lacking self-confidence
- lacking positive body image
- rebelling against perceived over-protectiveness of parents
- rebelling against excessive dependence upon others for assistance
- refusing to take prescribed medication
- teasing by peers
- difficulty making friends
- dating problems
- dressing/undressing inhibitions
- difficulty functioning in a regular class setting
- acting out of feelings of anger and hostility
- fears regarding impending surgery
- embarrassment over post surgery scars
- fearing the occurrence or recurrence of spontaneous lung collapse
- fearing heart attack
- fearing sudden death

Through a carefully planned and developed program of physical activity, designed specifically to meet the special needs of the affected child, psychosocial problems can be addressed. Based upon the individual's abilities and needs, and developed to insure the participant's enjoyment and success, such a program has the potential to enhance the psychosocial and physical well-being of the child with Marfan syndrome.

Psychosocial Goals

In addition to physical goals, psychosocial goals are a priority when developing an appropriate and effective physical education program for children with Marfan syndrome. Psychosocial goals must be developed to address the child's feeling different, handicapped, unattractive, humiliated, depressed, or embarrassed, to name just a few. Enjoyment, success, and positive interaction with peers in a well-planned physical activity program can improve self-esteem, confidence, and body image while reducing fears and inhibitions.

The attainment of established psychosocial goals is an important part of any educational experience. However, the attainment of these goals is especially important to handicapped children who are confronted with additional problems, while attempting to cope with the difficulties normally associated with growth, adolescence, and puberty. For children with Marfan, peak periods of psychosocial difficulty appear to occur during adolescence and during the time of a surgical procedure.

Based upon the physical characteristics and their psychosocial implications, potential psychosocial goals for these children are as follows:

- Increase the child's self-confidence
- Improve the child's body image
- Provide the child with opportunities for success and enjoyment through physical activity
- Improve the child's feelings of self-worth and self-esteem
- Improve the child's feelings of masculinity or femininity
- Increase the child's feelings of independence
- Increase the child's acceptance of others
- Promote the child's mastery of relaxation techniques
- Increase the child's knowledge and understanding of all aspects of the Marfan syndrome
- Teach the child to cope with peer pressure
- Educate the child's peers regarding all aspects of the Marfan syndrome
- Assist the child in recognizing anger, its causes, and acceptable methods of expression.
- Provide support related to all aspects of surgery.

Chapter 5

CURRICULUM

Goal

- To provide the reader with information regarding the selection of appropriate activities for inclusion in an adapted physical education curriculum for children with Marfan syndrome.

Objectives

- To provide the reader with information regarding individualization of the program, categorization of physical activity, suggestions for modifying activities and promoting outcomes, and factors to be considered in selecting or rejecting curricular activities.

- To provide the reader with curriculum recommendations, including activities that are approved and those that should be considered for adaptation or contraindication.

Individualization of Program

According to Dr. Reed Pyeritz, one of the world's leading Marfan syndrome experts, "In many cases, the potentially serious consequences of inappropriate physical activity for children with Marfan syndrome are offset by the positive benefits available through appropriately planned physical activity, and participation by these children should be encouraged." Given this endorsement, the curriculum developed for children with Marfan should be based upon carefully selected and planned physical activity which remains within the parameters permitted by medical professionals.

In prescribing physical activity for children with special needs, medical professionals and physical educators must treat each child as a unique individual. Parameters for exercise, and the selection of specific activities, must be determined on the basis of the child's needs, abilities, and limitations. Physical, emotional, psychological, and psychosocial factors must be addressed and consideration should be given to cognitive, affective, and psychomotor outcomes.

Although some general guidelines can be established for these children, the variability of the disorder requires that specific decisions be made on an individual basis. For example, if there is no evidence of aortic dilatation, and if there is ongoing echocardiographic review, the physician may permit participation in activities or exertion at higher levels of intensity. Such a decision rests solely in the hands of medical professionals.

Individualization of an adapted physical education program is accomplished within the "Individualized Education Program" (IEP) developed for the handicapped child. The IEP is developed with input from those involved in the child's education, including the child, parents, teachers, other school personnel, and physicians. Ongoing review of the IEP is required.

Categorization of Physical Activity

Physical activities may be categorized in many different ways. While there are significant limitations to such classification, it can assist in the consideration of appropriateness of physical activity for children with Marfan syndrome. For example, activities may be categorized in terms of their intensity or exertion levels. They may be categorized according to the potential for bodily contact and collision or on the basis of effect upon the body, that is, "static" or "dynamic" in nature. Still another category refers to an activity's "life-time" or "carry-over" value, which refers to its potential usage throughout life, especially as recreational exercise after formal school years. Classification according to activity type, such as games, aquatics, dance, gymnastics, and others, is also commonly utilized.

In terms of intensity, activities may be further broken down into light, moderate, or vigorous categories. Most activities, however, may be classified within all three of these categories, depending upon the level of exertion and intensity at which the activity is performed (e.g., jog, run at moderate pace, or run "all out"). Adaptation of activities according to implements used, space, frequency, intensity, and duration, can dramatically change the nature of exercise. Aerobic and anaerobic processes apply to the intensity of physical or muscular activity and the presence of adequate oxygen for the body to oxidize its carbohydrate sources of energy. In general, aerobic activity describes exercises that are low enough in intensity to be maintained for at least five minutes or longer. Anaerobic activity is so intense that within two minutes or less, the individual is exhausted. Energy for such activity basically occurs without the benefit of oxygen, since the demand is so great that the body cannot effectively transport an adequate supply rapidly enough through the lungs and cardiovascular system.

Generally, exercise can be divided into two types: static and dynamic. Dynamic, or isotonic exercise, involves changes in muscle length, joint movement, and rhythmic contractions which develop a relatively smaller force than during isometric exercise. When performed with a large muscle mass, dynamic exercise causes marked increases in both

oxygen consumption and cardiac output. During this type of exercise, there is an increase in stroke volume and associated minor changes in mean arterial pressure .

Static, or isometric exercise, involves little or no change in muscle length or joint movement and development of a relatively larger force. Static exercise usually involves a smaller muscle mass than dynamic exercise and causes a lower increase in oxygen consumption and cardiac output. Stroke volume changes little, and there is an increase in mean arterial pressure. This type of exercise, however, produces an increase in the contractile state of the left ventricle of the heart and a resultant pressure load.

Most physical activities have both dynamic and static aspects, and these categories simply serve to represent two extremes on a continuum. Swimming, which involves principally dynamic demands, and weight lifting, which involves principally static demands, each include elements of the other type.

In some cases, a "graded" exercise program is recommended by physicians to increase the efficiency of the cardiorespiratory system in heart patients. In such instances, an evaluation of the individual's exercise tolerance is made to determine the functional capacity of the heart and its reaction to exercise. The number of pulse beats per minute serve as an indicator of the functional capacity of the heart, with the amount of work that the individual can perform being expressed in a unit known as a "MET" or metabolic equivalent. One MET, or the amount of oxygen the body requires while sitting quietly, is equal to 3.5 milliliters of oxygen per kilogram of body weight per minute. The work metabolic rate is divided by the resting metabolic rate to determine the METs expended during physical activity. In this regard, it is normally recommended that a heart patient work at 60% to 70% of maximum functional capacity; in Marfan syndrome patients, a limit of 50-60% of maximal heart rate, at most, is appropriate. When supplied with this information, the physician and physical educator can refer to physical activity charts, available through the American College of Sports Medicine, which identify the energy demands of various activities.

Activities may also be categorized according to their potential for contact or collision. Such potential contact may occur with other participants, physical objects in the area, or implements used in the activity.

Suggestions for Modifying Activities and Promoting Outcomes

Listed below are suggestions for modifying activities and promoting outcomes for inclusion in an adapted physical education program for children with Marfan syndrome.

These adaptations and considerations are intended to minimize or eliminate aspects of physical activity which are inappropriate and to insure the achievement of desired outcomes. These include the following:

To address intensity, endurance and fatigue

- adjust duration of an activity
- adjust size of playing area
- use "quiet" or board games such as checkers, chess, or "Nok-hockey"
- use frequent "time out" periods
- permit participation at the child's own rate, with freedom to rest as necessary
- eliminate competitive and emotional stress factors
- reduce weight of implement
- use transportation or support devices

To address collision and contact concerns

- assign "zones" of play
- use individual activities
- use "singles" rather than "doubles" in racquet games, where appropriate
- change nature of implement (utilize "Nerf," foam, and "rag," items)
- group children according to size, abilities, and needs
- provide clear and concise directions, rules, and regulations
- provide play area free of obstacles, barriers, or hazards
- insure proper padding of facilities and equipment according to activity

To address visual and perceptual motor limitations

- change implement to decrease speed of flight and movement (e.g., a whiffle ball instead of a softball)
- place child to insure a clear and close visual field
- increase the size of the implement when necessary
- use brightly colored implements, lines, targets, and boundary markers
- provide playing areas that are free of hazards
- use Velcro to assist in "catching"

To address psychosocial factors and outcomes

- minimize stress and competitiveness in activity environment
- use music and relaxation techniques
- promote the worth and dignity of all persons and respect for individual differences during participation in physical activities

- select activities which promote self-esteem and feelings of self-worth
- select activities which will promote continued participation in physical activity and enjoyment on the part of the child
- select activities which are challenging, yet realistic, and which will result in success for the participant

It is important to note that adapting or modifying activities should not create an advantage to one individual over another. To retain the originality of an activity, minimize changes without sacrificing appropriateness for the child.

Factors in Selecting or Rejecting Curricular Activities

When selecting or rejecting curricular activities in an adapted physical education program for children with Marfan syndrome, consideration should include the following:

- The physician should work with the child, the parents, and school personnel in developing an appropriate exercise program in lieu of standard physical education.

- Experts believe that contact sports and those requiring sudden exertion should be avoided. A blow to the chest or the strain of jumping or stretching can result in serious injury. The larger the aortic root dimension, the greater the chance that sudden decelerations, like those that occur in body contact sports, can cause aortic dissection.

- Almost all physical activity involves some aspect of body collision if not presented safely. However, some require additional consideration, for handicapped or non-handicapped individuals, simply because of the dangers inherent in the activity itself (e.g., archery, bowling, golf, fencing, riflery, water activities, ice activities, and snow activities).

- Exercise at maximal capacity should be prevented to avoid the increases in heart rate, blood pressure and force of muscular contractions that impose stress on the aorta.

- The child with Marfan syndrome should be encouraged to take part in noncompetitive activities which can be performed at a reduced level of effort to control heart rate.

- Muscular underdevelopment and joint laxity will limit the physical activity of some children.

- The cardiologist should restrict physical activity further when aortic dilatation becomes appreciable (50 mm in adults). A cardiovascular surgeon should be contacted when aortic size approaches the threshold recommended for surgery (55-60 mm in adults).

- Physical therapy and muscle strengthening are useful approaches when working with individuals who demonstrate joint hyperextensibility.
- Activities which may rupture lung bullae, such as contact sports and decompression activities, should be contraindicated, including diving and scuba diving.
- Isometric exercises, weight lifting or training, and participation in other forms of physical activity to the point of exhaustion are contraindicated.
- Some activities may be proscribed because of their impact upon the joints, ligaments, and muscles. Examples include trampoline, vaulting, and gymnastic activities.
- The humanistic approach, which considers the human dignity and self-worth of all persons, should be applied when working with all children.
- An environment in which peer pressure frustrates the best intentions of the child's activity limitations is inappropriate and unacceptable.
- Athletes with a definite diagnosis of Marfan syndrome should be prohibited from participating in vigorous, competitive sports.

Curriculum Recommendations
for an Adapted Physical Education Program
for Children with Marfan Syndrome in Grades K - 12

Activity	Generally Approved	Activities which may require Adaptation or Contraindication
Aquatics/Water Activities	Drown-proofing skills safety skills swimming strokes	crew/rowing, diving, fishing, life-saving, sailing, scuba diving, snorkeling, water polo, water skiing
Dance/Rhythms	Rhythmic elements, singing games, folk, square, social	Ballet, cheerleading, jazz, kick-line, modern, tap
Games of Low Organization Grades K-4	Selected Games*	
Team Games Grades 3-12		Baseball, basketball, field hockey, football (all), ice hockey, lacrosse, polo, rugby, soccer, team handball, volleyball
Gymnastics/Tumbling	Balance	All other

Curriculum Recommendations (cont'd)

Activity	Generally Approved	Activities which may require Adaptation or Contraindication
Individual/Dual Activities Grades 3-12	archery, billiards/pool, board games, bowling (all), bicycling (stationary and/or leisurely) croquet, darts, golf, horseshoes, relaxation exercises, riflery, shuffle board, walking.	aerobics (all), badminton, boxing, curling, cycling, "duck walking," equestrian (all), fencing, handball, ice skating (all), isometric exercise, martial arts (all), mountain climbing, orienteering/back packing, paddleball, physical fitness testing, race walking, racquetball, roller skating, rope jumping, rowing/canoeing/kayaking, running (all), snow skiing (all), snow sledding (all), snowshoeing, squash, stair/stepping exercises, table tennis, track and field events, "tug of war," ultimate frisbee, weight lifting, wrestling
Movement Exploration Grades K-3	Selected Activities*	

* Activities in the games of "low organization" and "movement exploration" categories must be evaluated for appropriateness on the basis of the exercise parameters permitted and then selected or rejected accordingly.

"All" indicates that generally all types and variations of that activity are approved or contraindicated as listed. For example, "snow skiing" includes downhill, slalom, cross country, and ski jumping.

In the "team games" category, some skills within the activities may be approved.

Chapter 6
SUMMARY

This manual was written to serve as a primary resource for those involved in developing appropriate physical education experiences for children with Marfan syndrome. By providing information regarding the nature of the disorder, physical exercise guidelines, and curriculum activity suggestions, it is hoped that misinformation and associated fears will be eliminated or minimized.

Each child must be treated as an individual with unique abilities and needs. The safety and well-being of the child should be given the highest priority by all involved in the child's education and medical care. The physical education of a child with Marfan syndrome must be a joint venture among the child, parents, educators, and medical professionals. It is in the best interest of the child that there be open and ongoing communication among involved individuals.

The final decision pertaining to physical exercise, and the parameters within which such activity may take place, must necessarily rest in the hands of medical professionals. Once parameters, goals, and objectives have been established, however, it is the responsibility of the physical educator to develop a physical activity program designed to achieve the desired outcomes while remaining within the established bounds. In addition to determining the physical activities to be included in such a program, the physical educator must also address the associated psychosocial needs of the child with Marfan syndrome.

Some feel that placement in an adapted physical education program, or classification by the IEP committee, stigmatizes or negatively "labels" the child. However, the child afflicted with the Marfan syndrome has special needs which cannot be met safely or effectively in regular physical education programs. Public Law 94-142, and its subsequent revisions, was written in response to public outcry for guarantee to handicapped children that which is automatically given to every non-handicapped child: a free and appropriate education. Under the mandates of the law, an individualized education program (IEP), set in the least restrictive environment, will be developed to meet a Marfan child's special needs. For the child with Marfan syndrome, the alternatives to such placement — participation in the regular class program, serving as a "scorekeeper," or complete prohibition from participation — are inappropriate, unacceptable, and potentially unsafe.

The physical educator and physician, along with the child and parents, play a crucial role in establishing a positive school and learning environment for the child. The adapted physical educator and physician must be advocates for the handicapped, and work to provide an atmosphere built upon trust, support, and caring. Such a climate promotes feelings of positive self-esteem, helping the child to feel more a part of the school, and less "different." For the child with Marfan syndrome, the positive benefits of formal classification by the CSE, including the provision of appropriate physical education experiences, far outweigh the potential negatives.

A heightened awareness of the Marfan syndrome on the parts of educators and medical professionals helps to provide a physical education experience that is safe, effective, and appropriate. It is hoped that this manual will be of use to physical educators, coaches, and physicians in creating a physical education program that meets the special needs of these special children.

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Organizational Resources

For genetic information, or genetic counseling referrals:

Alliance of Genetic Support Groups
1001 22nd Street, NW
P.O. Box 800
Washington, DC 20037
(800) 336-GENE or (202) 331-0942

March of Dimes Birth Defects Foundation
1275 Mamaroneck Avenue
White Plains, N.Y. 10505
(914) 428-7100

National Society of Genetic Counselors
233 Canterbury Drive
Worthingford, PA 19086
(215) 872-7608

NIH/National Center for Education in Maternal and Child Health
(NCEMCH)
38th & R Streets, NW
Washington, DC 20057
(202) 625-8400

National Organization for Rare Disorders (NORD)
P.O. Box 8923
New Fairfield, CT 06812
(203) 746-6518 or 1-800-999-6673

For information about the Marfan syndrome:

National Arthritis and Musculoskeletal and Skin
Diseases Information Clearinghouse
Box AMS
Bethesda, MD 20892
(301) 468-3235

National Health Information Clearing House
P.O. Box 1133
Washington, DC 20013
(800) 336-4797

National Marfan Foundation
382 Main Street
Port Washington, N.Y. 11050
(516) 883-8712

Glossary

Adapted Physical Education refers to the modification of traditional physical activities to enable individuals with handicaps to have the opportunity to participate safely, successfully, and with satisfaction.

Aerobic Muscle Activity is generally exercise that is low enough in intensity to be carried on for at least five minutes or longer. The metabolic processes that supply the energy needs of muscle contraction during such activity normally take place in the presence of adequate oxygen to oxidize completely the carbohydrate sources of energy to carbon dioxide and water.

Ambloypia is impairment of vision without detectable organic lesion of the eye.

Anerobic Muscle Activity is exercise that is so intense that exhaustion ensues within one to two minutes or less, and for which energy must be supplied by anaerobic processes (without oxygen) because the oxygen cannot be transported via the lungs and cardiovascular system rapidly enough to meet such a demand.

Aortic Regurgitation is the backflow of blood from the aorta into the left ventricle due to the imperfect functioning of the aortic semilunar valve.

Aphakic Refraction is the the determination of the refractive errors of the eye and their correction by eyeglasses in the absence of the lens of the eye.

Arachnodactyly is a condition characterized by abnormal length and slenderness of the fingers and toes. In the past it was a synonym for Marfan syndrome.

Autosomal-Dominant Disorder is a disorder in which males and females are equally affected. A person with such a disorder has, at each conception, a 50% chance of passing the condition on to a child.

Bentall Procedure is a surgical procedure used to repair an aneurysm of the ascending aorta in Marfan patients. The procedure involves the use of a composite graft to replace both the aortic valve and the area of the aorta containing the aneurysm.

Beta-Adrenergic Blocking Agents are medications which decrease cardiac rate and output and reduce blood pressure. Propranolol is one such agent.

Chromosomes are structures in the nuclei of animal cells containing linear threads of DNA. They transmit genetic information and are associated with RNA and histones; 46 are normally present in humans.

Collagen is a protein substance of the white fibers of skin, tendon, bone, cartilage, and all other connective tissue.

Contractural Arachnodactyly is contractures of long, thin, spider-like fingers and toes.

Cystic Fibrosis is a generalized, autosomal recessive disorder of infants, children, and young adults in which there is a widespread dysfunction of the exocrine glands. Cystic Fibrosis is characterized by signs of chronic pulmonary disease due to excess mucus production in the respiratory tract.

Dilatation of the Aorta is a widening of the aorta.

Dissection of the Aorta is a tear in the inner and middle layers of the aortic wall resulting in the separation of these layers and the creation of a second channel through which blood is pumped. The force exerted by the blood pumping through the second channel may eventually rupture the outer wall of the aorta.

Dolichostenomelia is a synonym for arachnodactyly.

Dural Ectasia is dilatation, expansion, or distention of the dura mater covering the spinal cord.

Dynamic Exercise is "isotonic" exercise which involves changes in muscle length, joint movement, and rhythmic contractions which develop a relatively smaller force than during isometric exercise. When performed with a large muscle mass, dynamic exercise causes marked increased in both oxygen consumption and cardiac output. During this type of

exercise, there is an increase in stroke volume and associated minor changes in mean arterial pressure.

Dystrophia Medosdermalis Congenita is a synonym for Weill-Marchesani syndrome, a congenital disorder of the connective tissue transmitted as an autosomal dominant or recessive trait. It is characterized by short stature with a broad chest and heavy musculature, reduced joint mobility, ectopia lentis, myopia, and glaucoma, among others.

Ectopia Lentis is subluxation (dislocation) of the eye lens.

Ehlers-Danlos Syndrome, also called "cutis elastica" or "hyperelastica," is a group of inherited disorders of the connective tissue, varying in severity from mild to lethal, and transmitted genetically. The major manifestations include hyperextensible skin and joints, easy bruisability, friability of tissues with bleeding and poor wound healing, calcified subcutaneous spheroids and pseudotumors, and gastrointestinal, orthopedic, and ocular defects.

Elastin is a protein and an essential constituent of elastin fibers. It is brittle when dry, but flexible and elastic when moist.

Emphysema is the pathological accumulation of air in tissues or organs, especially the lungs.

Fibrillin is a structural protein; the primary constituent of microfibrils, which combine with elastin to form elastic fibers. Mutations in the gene for fibrillin cause the Marfan syndrome.

Fibronectin is an adhesive glycoprotein. One form of fibronectin is a cell-surface protein which mediates cellular adhesive interactions. It is important in connective tissues and cross-links to collagen.

Genes are segments of DNA molecules, the biological unit of heredity. They are self-reproductive and transmitted from parent to progeny.

Genetic Heterogeneity is the production of identical or similar phenotypes by different genetic mechanisms.

Genu Recurvatum is hyperextension of the knee.

Hemophilia is a hemorhagic diathesis occurring in two main forms and determined by a mutant gene. It is characterized by subcutaneous and intramuscular hemorrhages; bleeding from the mouth, gums lips and tongue; hematuria and hemarthroses.

Heritable Disorders are inherited as genetic traits.

Hernia is the protrusion of a loop or knuckle of an organ or tissue through an abnormal opening.

Homocystinuria is an autosomal recessive aminoacidopathy characterized by excessive homocysine in plasma and urine. Some manifestations (tall stature, scoliosis, pectus deformity, ectopia lentis) resemble the Marfan syndrome.

Hypermobility is excessive mobility, as in a joint.

Inguinal hernia is a hernia into the inguinal canal, either directly or indirectly.

Kyphoscoliosis is the backward and lateral curvature of the spinal column.

Lordosis is the anterior concavity in the curvature of the lumbar and cervical spine as viewed from the side. It is also referred to as "hollowback, saddleback, or swayback".

Marfan Syndrome is a congenital disorder of connective tissue characterized by abnormal length of the extremities, especially of fingers and toes, dislocation of the lens, cardiovascular abnormalities (commonly widening of the ascending aorta), and other deformities. It is an autosomal dominant disorder with variable degrees of expression.

Menarche is the establishment or beginning of the menstrual function.

MET is the amount of oxygen the body requires while sitting quietly. One MET equals 3.5 milliliters of oxygen per kilogram of body weight per minute.

Microfibril is a minute fiber or filament.

Mitral Regurgitation is the backflow of blood from the left ventricle into the left atrium due to the inadequate functioning of the mitral valve.

Mitral Valve Prolapse is the sinking or falling of the mitral valve leaflets.

Myopia is the error of refraction in which rays of light entering the eye parallel to the optic axis are brought to a focus in front of the retina because the eyeball is too long from front to back or because there is increased strength in refractive power of the media of the eye. It is also called nearsightedness.

Neuropsychological Deficit denotes a deficit of the nervous system and mind.

Pectus Carinatum is undue prominence of the sternum. It is also called "chicken or pigeon" breast.

Pectus Excavatum is undue depression of the sternum. It is also called "funnel" chest or breast.

Pes Planus is a deformity of the foot in which the position of the bones, relative to each other, has been altered, lowering the longitudinal arch. It is also called "flat feet".

Physical Education is a term denoting the development of the following: physical and motor fitness; fundamental motor skills and patterns; and skills in aquatics, dance, and individual and group games and sports (including intramural and lifetime sports). The term includes special physical education, adapted physical education, movement education, and motor development.

Pneumothorax is a collapse of a lung, usually due to rupture of a bleb on the surface of the lung that allows entry of air into the space between the lung and the chest wall.

Scoliosis is an appreciable lateral deviation in the normally straight vertical line of the spine.

Sickle Cell Disease is a hereditary, genetically determined, hemolytic anemia occurring almost exclusively in Blacks. The disease is characterized by arthralgia, acute attacks of abdominal pain, ulcerations of the lower extremities, sickle-shaped erythrocytes in the blood, and homozygous presence of S hemoglobin in the red blood cells.

Static Exercise is "isometric" and involves little or no change in muscle length or joint movement, or development of a relatively larger force. Static exercise usually involves a smaller muscle mass than dynamic exercise and causes a lower increase in oxygen consumption and cardiac output. Stroke volume changes little and there is an increase in mean arterial pressure. Static exercise produces an increase in the contractile state of the left ventricle of the heart and a resultant pressure load.

Steinberg Thumb Sign is the relatively narrow palm of the hand, long thumb, and longitudinal laxity of the hand. The thumb apposed across the palm extends well beyond the ulnar margin of the hand. It is a simple and non-specific test for the presence of the Marfan syndrome.

Striae Atrophicae are linear, depressed, atrophic, pinkish or purplish scar-like lesions that later become white as they mature. They appear on the abdomen, breasts, buttocks and thighs due to the weakening of the elastic tissues.

Subluxation of the Lens is a partial dislocation of the lens of the eye.

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About the Author

Thomas J. Romeo has been involved in the fields of physical education, coaching, and administration for over thirty years.

He is the director of athletics and the coordinator of physical education and health for the Port Washington, New York, public schools, and he has served as a consultant in adapted physical education to neighboring school districts in New York and New Jersey.

This manual is the culmination of Thomas Romeo's doctoral project at New York University, and for this work he was awarded his degree.

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ERRATA

Page 7, Line 15 from bottom should read: "other health impaired."

Page 20, Line 16 from top should read: appropriateness and/or

Page 38, Line 9 from bottom under "Sensitivities/Competencies Needed by Physical Educator" should read : appropriate physical

Page 46, Line 4 from top under "Sensitivities/Competencies Needed by Physical Educator" should read: visual needs and the ability to use developmental and/or remedial approaches

Page 47, Line 10 from bottom under "Instructional Methods & Strategies" should read: fears and information

Page 64, Line 9 from top should read: **Amblyopia** is

Page 67, Line 11 from top should read: homocystine in plasma and urine.